



# **CARDIAC CATHETERIZATION IN CONGENITAL HEART DISEASE**

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# CARDIAC CATHETERIZATION IN CONGENITAL HEART DISEASE

A CLINICAL AND PHYSIOLOGICAL STUDY  
IN INFANTS AND CHILDREN

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To the memory of  
Pierre Birel Rosset Cournand  
*"Having fulfilled his course in a short time,  
he fulfilled long years "*



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## INTRODUCTION

IN recent years the technique of right heart catheterization has been increasingly applied to the diagnosis of congenital malformations of the heart and large vessels. In 1945-46 Brannon, Weens, and Warren reported the use of this technique in the diagnosis of auricular septal defects, and E. de F. Baldwin, Moore, and Noble reported its use in the diagnosis of interventricular defects. Since then two other groups working under the direction of Dexter and of Bing, have reported their findings in various types of congenital cardiac malformations and have described in detail the procedure followed in their studies. The contribution of both groups has been outstanding and their published papers should be read by anyone interested in this field of investigation.\*

So far, however, there has been no concerted effort by clinicians and investigators to pool the results of their separate studies, to compare and to integrate clinical and physiological data. This monograph is presented as an attempt in such a direction.

A study of a varied group of congenital defects of the heart was begun by the present authors in January 1946 as a joint undertaking between the Pediatric Service and the Children's Cardiac Clinic of Bellevue Hospital (New York University Division), the Children's Cardiac Clinic of Lenox Hill Hospital, and the Cardio Pulmonary Laboratory of the Chest Service of Bellevue Hospital (Columbia University Division). One of the participants was especially interested in pediatrics and cardiology, while the others had had experience with the technique of cardiac catheterization in problems related to the physiology of the circulation in adults. In the present investigation it was felt particularly important to explore the possibility of extending the method of cardiac catheterization to the study of young children and infants. But in order to achieve this it was found necessary to introduce two new features. Because of the small size of the arm vein in many cases, the saphenous vein at the femoral region was chosen as the site for introducing the catheter into the venous system. Also, owing to lack of voluntary cooperation in the younger age group and the necessity of maintaining a steady state throughout the period of study the use of general anesthesia was adopted. In view of the need of an anesthetic which would not interfere with gas analysis, avertin was tried and found satisfactory.

The essential purpose of a combined study was to make observations of a physiological nature in various types of congenital anomalies of the heart, and to compare and correlate these observations with physical signs and x-ray findings. It was expected to gain additional diagnostic information and a better understanding of the physiological factors underlying clinical observations as well as to add to the knowledge in the field of the physiology of the circulation.

*This monograph is divided into two parts. In the first part the methods used in the physiological studies are described and discussed. In the second part individual cases illustrating various types of malformation are presented.*

Some of this material was exhibited at the International Congress of Pediatrics held in New York City in July 1947. On this occasion all the data were presented in a simple schematic form and were correlated and compared. The method of presentation proved so helpful in demonstrating the respective values of various aspects of the clinical and physiological studies that it has been retained in this monograph. Several cases having been added to the original material. The cases were chosen in order to give a general idea of the most common problems we have encountered in this field of investigation.

In view of the fact that extensive studies of the cyanotic group are being carried out by Bing at the Johns Hopkins School of Medicine the emphasis here was deliberately placed on the other types of malformation of the heart. Surgical and pathological confirmation of the diagnosis has

\* See Bibliography, pp. 107-108.

been available only in certain of the cases studied. We therefore consider that the presentation we are making at this time, with the above limitations, is in the nature of a pilot endeavor. It is hoped that the results of other investigations carried out along similar lines in a greater number and larger variety of cases, and with surgical or pathological data, will be made available to those interested in this group of fascinating but often baffling cardiac conditions.

We wish to acknowledge our indebtedness to Dr. Hurley L. Motley, who analyzed most of the blood pressure tracings and mounted them for reproduction; to Dr. Lars Werkö, Dr. David T. Dresdale, and Dr. Herbert M. Weiner for their contribution in recording the blood pressure tracings; to Mrs. Marianne S. Lester, Miss Blanche Spierto, and Miss Dorothy Allen, whose accuracy and efficiency in the laboratory work were of invaluable help; to Dr. E. A. Rovenstine, Dr. Emmanuel M. Papper, and other members of the Department of Anesthesia of Bellevue Hospital; to Dr. Charles W. Lester and Dr. Gertrude B. Nicolson, who referred cases to us for study; to Miss Rae Morse, who prepared the manuscript; to Dr. Réjane Harvey and Dr. Irené Ferrer, who read it; to Dr. Domingo Gomez for his many helpful suggestions; to Dr. L. Emmett Holt and Dr. J. Burns Amberson, chiefs respectively of the Pediatric Service and the Chest Service of Bellevue Hospital; to Dr. Dickinson W. Richards, Jr., for his inspiring leadership and continuous encouragement; and finally to the Commonwealth Fund, which has supported the Cardio-Pulmonary Laboratory for many years and which undertook the publication of this monograph.

A. C.  
J. S. B.  
A. H.

*October 1948*

## PART ONE

## THE PHYSIOLOGICAL METHODS

THE physiological methods of the present study are based on the following three techniques (1) right heart catheterization (2) arterial puncture, and (3) collection of expired air for measuring the oxygen consumption. All the measurements were made in the recumbent position and in the post absorptive state. Occasionally a mild degree of physical activity was induced.

The technique of right heart catheterization affords a means of sampling mixed venous blood in the right heart and the pulmonary artery. In a given subject maintained in a steady state variations in oxygen content of blood samples taken in rapid succession from the right auricle, the right ventricle, the pulmonary artery, and its two main branches are small. Abnormal communications between these structures and the pulmonary vein, the left auricle, the left ventricle, or the aorta result in contamination of mixed venous blood with highly oxygenated blood, as the pressure conditions prevailing in the circulation favor blood shunts directed from the left cavities and their dependent vessels to the right cavities and the pulmonary artery. Therefore the approximate location of the shunts can be determined by gas analysis, and the volume of blood shunting can be calculated. Moreover, the presence of an abnormal communication may be revealed by the unusual routes eventually taken by the tip of the catheter as it is introduced into the heart. Contamination of pulmonary venous blood by mixed venous blood can also be recognized by arterial blood gas analysis. Such contamination occurs when the dynamic conditions prevailing in the heart in the presence of a cardiac malformation favor a shunt directed from the right cardiac cavities into the left cardiac cavities or the aorta.

With simultaneous determination of the oxygen intake in the lungs and of the oxygen content in the heart and in a systemic artery, the calculation of blood flow in the systemic and pulmonary circuits and of blood shunts through abnormal communication becomes a matter of applying simple formulae.

In addition, the two techniques of right heart catheterization and arterial puncture provide a means of recording blood pressures. The patterns of the curves and the range of normal pressure variations in the right or left auricle, the right or left ventricle, the pulmonary artery, and the aorta and peripheral arteries are well defined. Hence blood pressure recordings may serve two purposes: (1) identification of the cardiac cavities or large vessels in which the tip of the catheter is placed; (2) study of the effects of cardiac malformations upon the dynamics of the circulation.

A well trained team and a well organized laboratory are essential for the successful application of the physiological methods. The reliability of the results and the safety of the method depend upon meeting both these requirements.

The team should consist of three physicians working as an integrated unit, but each responsible for his part in the procedure and each constantly on the alert for changes in the patient's physiological status and for technical errors. One member of the team, thoroughly familiar with every aspect of the techniques and of the procedure, should be in charge of the study, so that the various steps may follow with a minimum of delay. He directs the procedure and interrupts it immediately if he deems it advisable. The laboratory should be organized with a group of technicians well trained in the rapid and accurate analysis of respiratory gases in air and in blood samples.

The material presented in the following chapters may be of some help to investigators who are planning similar physiological studies. It is the opinion of the authors, however, that a prolonged period of training with a team in a laboratory already in operation is the best means of acquiring a thorough knowledge of the method, including its advantages as well as its pitfalls.

Chapter I describes the equipment, the techniques, and the procedure employed in the course of the physiological studies and discusses the precautions to be observed in order to avoid undue risks and serious complications.

Chapter II illustrates the various appearances of the catheter inside the heart and the large vessels, as shown by fluoroscopy.

Chapter III demonstrates the characteristic patterns of the blood pressure tracings, as they are recorded

Chapter IV presents the formulae used in connection with these studies for the calculation of the blood flow in the systemic or pulmonary artery and the intra and extracardiac blood shunts

## EQUIPMENT

The equipment used in the physiological studies includes venous catheters indwelling arterial needles apparatus for measuring oxygen consumption and for recording blood pressures an instrument set up for the introduction of the catheter and the needle and finally a large number of syringes for the collection of blood samples

**Venous catheter** The catheter\* recommended is a modified ureteral type made of nylon with a smooth unwettable plastic covering flexible and radio opaque It is 100 cm long with only one opening at the tip and an airtight adaptor at the proximal end For convenience in manipulation a slight curve is provided in the distal 11 cm In the case of small children and infants a modified type with the curve starting 3 cm from the tip has been found useful A #6F or #7F catheter is preferable since pressure tracings are more satisfactory with these than with larger sized catheters and as a rule the withdrawal of blood samples is easy Sterilization is by boiling for 10 minutes in distilled water the catheter being then wrapped in a towel It should be sterilized some time before use because for a long period after heating it remains too flexible for easy manipulation Before introduction the catheter should always be carefully tested for surface imperfections and for leaks at the junction with the adaptor Immediately after use it should be soaked with tap water for several minutes then filled with Hemo sol<sup>†</sup> solution and allowed to soak in the same solution for 1 hour then rinsed again for 1 hour with tap water through a pressure connection These precautions are indispensable in preventing the deposition of particles of blood on the inner side of the woven fabric which might initiate a pyrogenic reaction in the course of the next catheterization

**Indwelling arterial needle** The needle† used for insertion in an artery is a modified Lindemann type in which a medium bevel #22 needle fits into a #19 outer sleeve with blunt tip The syringe adaptor is soldered about 1.5 cm down the #19 shaft the base of the shaft being allowed to project past the adaptor When a Luer lock syringe is attached for sampling this free segment goes directly into the end of the syringe and contamination from blood and blood clot lying in the fitting is thus avoided In the case of infants it is occasionally impossible to introduce an indwelling needle In this event a #20 needle with a short bevel is used for blood sampling and pressure recording but is not left in place

**Instrument set up** A sterile tray or truck is set up to carry the instruments and various sterile items required for skin incision isolation of the vein and closure of the incision They consist of two sheets and two large towels two towel clamps two small curved mosquito hemostats, two small straight mosquito hemostats a plain forceps a scalpel with a #11 blade a small scissors, a medicine glass for the novocain solution two #26 needles for local anesthesia two 5 cc. syringes a curved cutting edge needle with black silk for closure of the incision when necessary a small roll of black silk #0000 for ties several 3 way stopcocks and extra adaptors a large beaker to hold saline solution a 50 cc Luer lock adaptor syringe for testing the catheters for leaks a tourniquet Sterile gloves gowns sheets and infusion sets should be available

\* Manufactured by the U.S. Catheter and Instrument Corporation, Glens Falls, New York

† Manufactured by Becton Dickinson, Rutherford, New Jersey

A second tray holds from fifteen to twenty 20 cc. syringes with Luer lock adaptors to be used in the sampling of blood. These dry syringes are prepared by the introduction of a small globule of mercury, which fills the space between the rim of the plunger and the barrel and which is to be used in mixing the blood sample. The sides of the syringe are then moistened with 6 drops of the following solution: 600 mgm of sodium fluoride and 100 mgm of dried heparin in 15 cc of an 0.85 per cent sodium chloride solution. The excess is expelled through the aperture of the syringe held in upright position, the amount of solution left being too small to dilute the blood sample significantly. If the fit of the plunger is not perfect, as sometimes happens, its surface should be lightly greased with "Lubri-Seal" to avoid leaks and accumulation of air bubbles during sampling.

**Apparatus for the measurement of oxygen consumption.** The oxygen consumption may be determined by means of a standard metabolism apparatus, or by collecting samples of expired air in a Tissot spirometer or a Douglas bag and analyzing the gaseous content. In the case of infants and young children it is recommended that an anesthesia mask be used rather than a mouth piece or nose clip to connect the apparatus to the subject. A special mask\* which has proved very satisfactory consists of a face piece of light weight plastic (moulded for form fitting) and a wide detachable cuff made of high grade inflatable rubber and built over a sponge rubber cushion.

**Apparatus for recording blood pressures.** For gross measurement of mean pressures an anaeroid Tyco's manometer may be connected by a piece of rubber tubing to the catheter or the arterial needle. A few cubic centimeters of saline solution introduced in the tubing in the part proximal to the catheter attachment, is used to minimize the chances of air embolism. A saline manometer of the type used for the clinical measurement of venous pressure may be substituted for the anaeroid. For accurate measurement and recording it is necessary to use fluid filled manometers of the Hamilton type or an electrical recording device. In our work we have employed a special multiple blood pressure recording apparatus, designed and constructed by the Cambridge Instrument Company according to our specifications. It consists of the following parts which are mounted on a mobile cabinet table: (1) an electrocardiograph with camera, string galvanometer, time marker, standardizing box, camera motor with 3 speed gear, (2) four fluid pressure Hamilton manometers with beryllium copper membranes of different sensitivities which can be connected through fluid filled lead tubing to the proximal end of the cardiac catheter and arterial needle, (3) a fluid system with two 3 way control valves and a pyrex bottle (2000 cc. capacity) the latter serving as a reservoir for a saline or sodium citrate solution and provided with a pressure bulb for applying pressure in the fluid system, (4) a calibrating mercury manometer and an air trap bottle, (5) an optical system for transmitting the deflection of the membranes to a recording camera with a light source and a reflecting mirror. The optical system was designed in such a manner that there is no parallax between the simultaneous tracings of pressure and the electrocardiogram.

## TECHNIQUES AND PROCEDURE

**Set up for study under fluoroscopic and electrocardiographic control.** The studies are carried out on a fluoroscopic table covered with a sponge rubber mattress. In order to avoid excessive radiation of the patient and of the members of the research team it is recommended that certain precautions be taken such as limitation of the field to be fluoroscoped, the use of lead sheets as a protection against stray radiation, and the use of lead aprons and of a timing device.

Throughout the entire period of study the electrocardiograph is connected with the subject and one member of the team follows the deflection of the string in Lead II in the reflecting mirror.

\* Manufactured by V. Ray Bennett, 8154 Beverly Boulevard, Los Angeles, California.



*for cardiac rate and irregularity, being particularly watchful during manipulation of the catheter within the heart*

**Anesthesia:** In the case of children up to approximately 9 years of age catheterization is usually done under basal avertin anesthesia, given by a competent anesthetist, in order to maintain a steady state while obtaining multiple blood samples. Premedication consists of scopolamine or atropine, occasionally with some barbitol derivative. Avertin is used in an initial dose of 90 to 110 mgm per kilogram of body weight and supplemented by doses of 20 to 40 mgm per kilogram if the anesthesia begins to wear off during the procedure. In our studies the total dose was usually 140 mgm per kilogram. For local anesthesia at the site of introduction of the catheter and of the arterial needle, a 2 per cent solution of novocain was used liberally.

If it is thought that the child will cooperate and remain quiet throughout the procedure, local anesthesia at the sites mentioned above may be used—especially in the case of children of the older age group. Occasionally an infant can be maintained in a quiet state without narcosis during the entire procedure. The decision whether local anesthesia alone will suffice should be left to the physician best acquainted with the child's or infant's reactions.

**Technique of introducing the arterial indwelling needle:** The special indwelling needle is placed in the brachial artery whenever possible. Otherwise it is introduced in the femoral artery. The site of the puncture and the deeper tissues are thoroughly infiltrated with a 2 per cent novocain solution. The index finger is then placed over the infiltrated site and pressure is applied over the artery so that it stays in a given position while the underlying tissues are anesthetized. It is advisable to infiltrate the tissues on either side of the artery as well as above and below the chosen route along which insertion of the arterial needle is to be made. When the area has been anesthetized, the arterial needle containing the patent stylet is held in one hand while the index finger of the other applies sufficient pressure over the artery to fix its position but not to obliterate its pulsations. The arterial needle is then inserted at a point just below the index finger and driven slowly through the skin and underlying tissues at an angle of about 45 degrees. As progress is made in the direction of the pulsating vessel, the thumb is removed from the patent stylet at intervals in order to ascertain whether or not the artery has been punctured. When puncture of the artery has been accomplished, blood will be seen to drip from the patent end of the stylet. The flange of the needle is then grasped with one hand, the patent stylet with the other. Coincident with the slow removal of the patent stylet, the arterial needle is "threaded" into the lumen of the vessel. When the stylet has been removed, blood should spurt from the end of the needle. The solid stylet is then placed in the needle, and stylet and needle together are threaded further into the vessel until a substantial portion of the needle lies within the lumen of the artery. As soon as the needle is in place it should be taped securely to the skin and the arm or leg immobilized. As a precaution against clotting in the needle, the solid stylet should be dipped in hydrogen peroxide before it is replaced. During blood sampling, the stylet should stand in the peroxide.

#### **Technique of venous catheterization:**

**Choice of the site for introducing the catheter into the venous system:** Initially, a survey of the arms is made with a tourniquet applied. If a vein going into the median basilic system can be felt or seen and is of sufficient size, it is selected as the site for introduction. The left side is preferred because manipulation of the tip of the catheter inside the heart appears to be easier from this side but the right may also be used. The choice of a vein draining into the cephalic system should be avoided since the acute angle at the subclavian vein can seldom be passed. In the case of small children, or of those in whom no satisfactory arm vein can be found, the saphenous vein at the femoral area is used.

## *Isolation of the vein*

**Arm vein** The arm is prepared by scrubbing with soap and water and then painting with tincture of merthiolate. Sterile drapes are applied, so that the antecubital area is exposed in a sterile field. The operator wears sterile gown and gloves.

The area over the vein is infiltrated with a 2 per cent novocain solution. A 5 mm incision is made parallel and just lateral to the course of the vein, which is exposed by blunt dissection with a small, curved hemostat and then elevated into the wound. Two #0000 silk ligatures are passed under the vein and the distal one tied with a knot. The other is left untied and used as a sling to control back bleeding. A transverse nick is made in the anterior wall of the vein with a #11 blade. The proximal lip of the incision in the vein is grasped with a fine, straight hemostat and the lumen held open with it. The catheter is advanced into the vein after flow from the saline reservoir connected with it has been regulated at a rate of about 20 to 30 drops per minute.

**Saphenous vein** The preparations are similar to those for the exposure of the arm vein. The femoral artery is palpated an inch below the inguinal ligament, and an area just medial to it is infiltrated with 2 per cent novocain solution. A 15 mm incision is made in the line of the vein, and the vein isolated by blunt dissection. In infants this is a vessel from 2 to 3 mm in diameter. The vein is then handled in the same way as an arm vein, the two ligatures being placed and the vein opened. When the catheter is introduced, it can be felt to dip into the femoral vein, it is then advanced easily into the iliac veins.

**Manipulation of the catheter under fluoroscopic control** After the catheter has been advanced 20 to 30 cm, further manipulation is done under fluoroscopic control. This consists in advancing, rotating, and withdrawing the catheter until its tip is placed in the desired location. When the catheter is introduced into an arm vein, some difficulty may occasionally be encountered in the axilla or the tip may be seen moving cephalad into the jugular vein. If this occurs, the catheter is withdrawn, rotated, and advanced again with the neck flexed to the side, the arm abducted, and a slight traction exerted upon it. If the catheter shows a repeated tendency to cross the midline into the opposite subclavian vein, advancing it at the time of inspiration will facilitate its entry into the superior vena cava. The catheter should never be advanced against resistance. Caution should be exercised to avoid buckling or the formation of a "U". When seen to be pointing in a desired direction, the catheter is advanced rapidly.

To reach the right auricle, the tip is advanced until it lies about 2 or 3 cm above the level of the diaphragm. To enter the right ventricle, the tip is rotated to point toward the left and then advanced. On one such effort it will cross the midline. Passage into the ventricle is checked by noting the pressure, with either a Hamilton or a Tyco's manometer. To enter the pulmonary conus, the catheter is carefully withdrawn and advanced, or rotated and advanced within the ventricle until the tip can be seen to point cephalad. It is then advanced into the pulmonary conus and main branches of the pulmonary artery. Occasionally in the case of infants, when the catheter has been introduced into the saphenous vein, a loop will form in the right auricle while an attempt is being made to advance the tip into the outflow tract of the right ventricle and the pulmonary conus. The modified catheter with the curve near the tip was designed to remedy this difficulty. As soon as the tip of the catheter has been placed inside the heart shadow, the saline flow from the reservoir may be reduced and adjusted to 5 to 10 drops per minute.

**Sequence of studies:** The general plan of study consists in collecting blood samples and recording blood pressures in the heart and large vessels in rapid sequence. In most instances the tip of the catheter is first advanced to one of the two main branches of the pulmonary artery, and

from there on withdrawn into the superior ventr cavity in successive stages during which blood samples are taken and pressures are recorded. However unusual routes may be followed by the tip in various types of cardiac malformations. In these cases fluoroscopic examination in various positions, study of the pattern of the blood pressure tracings and gas analysis of the blood samples are essential in order to infer the location of the tip of the catheter. In general blood samples and blood pressure tracings should be taken in quick succession in two communicating heart chambers or in a heart chamber and the communicating large vessel. All blood samples should be withdrawn while the subject is maintained in a steady metabolic state. If this steady state is interrupted some time should be allowed to elapse before resuming the collection of blood samples and the recording of blood pressures at the site where the last observations were made. In a few cases of interventricular septal defect the oxygen content of the right auricle may be identical with that of the right ventricle in the quiet resting state and the shunt can be demonstrated only if the child becomes somewhat active or is induced to cry.

If during the early part of the study the tip of the catheter has been introduced into a chamber of the left heart or into the pulmonary vein or the aorta blood samples and pressures should be taken immediately in these locations. Otherwise the following order for sampling blood and recording pressures is recommended because it permits a rapid uninterrupted collection of data and a minimum of x-ray exposure: (1) one or if possible both branches of the pulmonary artery, (2) the truncus of the pulmonary artery, (3) the outflow tract of the right ventricle, (4) the mid portion or the apex of the right ventricle, (5) the tricuspid area on the ventricular side, (6) the tricuspid area on the right auricular side, (7) the mid right auricle, (8) the superior ventr cavity, (9) the inferior ventr cavity near its opening in the right auricle. The blood sampling and pressure recording in the peripheral artery should be done simultaneously with auricular sampling and recording in the right ventricle and if the opportunity presents itself in the aorta. A protocol should be kept of the sequence of the study and a brief descriptive note of the exact location of the catheter should be dictated each time a physiological observation is made. Occasionally x-rays are taken in various positions when the location of the catheter cannot be easily identified or is unusual.

**Collection and handling of blood samples.** For the collection of blood samples the following procedure is recommended:

A syringe containing a few cubic centimeters of a sterile saline solution and no air bubbles is adapted to the 3 way stopcock connecting the catheter to the saline reservoir. Through proper manipulation of the stopcock and the syringe it is made certain that the circuit does not contain any air bubbles and that flow is free in the catheter. Then suction is applied gently until blood flows back easily in the syringe. *No forceful suction is used while sampling in the right ventricle since it will often cause ectopic beats.* Then the Luer lock syringe used for sampling and prepared as described above is adapted to the 3 way stopcock and a blood sample of 5 to 10 cc. obtained. The sampling syringe is then disconnected and the 3 way stopcock and the catheter are immediately flushed with several cubic centimeters of a saline solution and the continuous drip started again. Meanwhile the syringe with the blood sample is stoppered with a toothpick. One small bubble of air is oftentimes present. It can be expelled easily. If the sample contains several bubbles especially in the form of foam it is discarded. The syringe is rotated slowly while being taken to the laboratory. Blood is transferred immediately under pressure into Oswald Van Slyke pipette by way of a small ( $\approx 23$ ) gauge needle connected with the syringe and inserted into the tip of the pipette. A small piece of rubber pierced by the needle makes possible an airtight seal. The carbon dioxide and the oxygen content and the oxygen capacity are immediately measured in duplicate using the Van Slyke-Neill apparatus. The syringes containing the blood samples are

stored in an icebox for later drawing of samples if required. In this case a period of 10 minutes should be allowed for rotation of the syringe before transfer of blood to the pipette.

**Recording of blood pressures** The recording of blood pressure is done immediately before and after the collection of samples. The free end of the lead tubings connecting with the manometer is fixed near the patient at a level corresponding to the plane of the heart, 3 cm. below the angle of Louis for intracardiac recordings, or at the level of the peripheral artery for intrarterial pressure recordings. The free side arm of the 3 way stopcock connecting the catheter and the saline reservoir is flushed with fluid from the saline reservoir and then attached to the lead tubing. The movements of the light beam are observed in the reflecting mirror in order to determine if artefacts are present or if the tracings are damped, in which case the tip may be displaced or the catheter flushed with saline. A short recording is taken, the picture of the baseline being made at the beginning and at the end of each recording. The film is immediately developed in an adjoining darkroom at the end of each step in the study.

It is of great importance that the member of the research team in charge of the study should constantly be informed of the results of the gas analysis as they are obtained and be shown the tracings as they are developed. In this manner the conduct of the study will be better controlled and plans altered according to findings. As a rule, a continuous recording is made while the tip of the catheter is slowly moved from the region of the pulmonary artery into the outflow tract of the right ventricle under fluoroscopic control, and in cases in which a sudden systolic pressure increase is observed, the point where this occurs is carefully noted. This maneuver is essential in differentiating between stenosis at the pulmonary valve from stenosis of the infundibular area. If a difference of pressure has been noted between the pulmonary artery and the ventricle, attempts should be made to obtain repeated tracings while moving the catheter back and forth past the pulmonary valvular region.

**End of the procedure** The catheter is withdrawn slowly but steadily. If the arm vein was entered, the tie applied to the vein at the beginning of the procedure is removed. Pressure is used to control bleeding. A small flamed adhesive strip is used to close the skin defect and a sterile dressing applied. The arterial needle is withdrawn and immediate pressure on the vessel is maintained for at least 5 minutes and pulses below are carefully checked. At any evidence of peripheral swelling, pressure is reapplied. If the saphenous vein was used, it is tied near the confluence with the femoral vein, penicillin is placed in the wound, and the lips of the wound are sutured.

## COMPLICATIONS

Most complications observed during or after completion of cardiac catheterization are of a minor character. Strict observance of details of technique given in the previous sections should help to avoid these. It must always be kept in mind during the procedure that some risk is involved and that the safety of the patient dominates all other considerations. The following is a brief discussion of the various complications related to the technique.

**Air embolism** The accidental introduction of small air bubbles into the right cardiac cavities by way of the catheter is of no importance as long as there is no abnormal communication with the systemic circulation, because these bubbles are probably effectively filtered out in the pulmonary capillaries. In the presence of congenital malformations of the heart, associated with hemodynamic conditions favoring right to left blood shunts, air emboli may be thrown into the systemic circulation if precautions against the introduction of air bubbles into the catheter are

not taken Embolization of one or several cerebral arteries is one of the most serious hazards of using the technique in this type of defect. However, in his studies of several hundred cases of intracardiac communications, Bing suspected it in two cases only. In no instance did it result in more than transient neurological symptoms or signs.

**Venous and intracardiac thrombosis:** The trauma incident to the incision of the vein and the introduction of the catheter often causes a limited thrombosis. This usually extends less than 5 cm. upward from the site of the incision. In order to maintain a free blood flow in the main brachial vein it is therefore very important to choose as the site of introduction of the catheter a branch of the brachial venous system which is distal to and at a distance of at least 5 cm. from the last confluence of veins of this system. Potentially, catheterization of the saphenous vein offers a greater risk of thrombosis of larger veins—namely, the femoral, with the attendant danger of pulmonary emboli. In order to minimize this risk, the saphenous vein should be tied off as close to the femoral vein as possible at the end of the procedure.

Intracardiac thrombosis, associated with traumatic lesions of the endocardium, is another potential hazard to be reckoned with. Goodale, Lubin, Eckenhoff, Haskenschiel, Durlacher, Landing and Banfield have described lesions following the prolonged introduction of catheters into the hearts of dogs.\* With one exception, however, such lesions have not been revealed in the pathological material gathered by many groups of investigators using the technique of cardiac catheterization in man. Only one autopsy report, by Johnson, Wollin, and Ross, is suggestive of an endocardial lesion. In the case of a young child with polycythemia and a congenital cardiac defect, who died one month after catheterization, a thrombus was found in the right auricle, inferior vena cava and iliac and renal veins. It is not clear from the description given whether the thrombus originated at the site of introduction of the catheter in the saphenous vein, or at the base of a leaflet of the tricuspid valve to which it was attached, or in both locations. Whether the hazard of venous thrombosis in patients with polycythemia is serious enough to warrant prophylactic anticoagulant therapy after catheterization is an important question. Because of our limited experience with this group, we are not in a position to give a documented answer.

**Rhythm disturbances:** The development of rhythm disturbances during cardiac catheterization is by far the most important complication to consider.

Premature contractions are often observed during the manipulation of the catheter inside the heart. These ectopic beats usually occur when the tip of the catheter is being passed through the tricuspid valve, or lies against the ventricular septum in the region of the outflow tract, or when suction is applied in sampling blood in the right ventricle. Ectopic beats are easily identified by watching the deflection of the galvanometer string. Changing the position of the catheter causes these beats to disappear.

Runs of ventricular tachycardia have been encountered on a very few occasions when the cause of abnormal stimulation was not removed. In our studies of congenital malformations of the heart we observed one patient with an interventricular septal defect in whom the introduction of the tip of the catheter through the defect stimulated many ectopic beats of bizarre electrical configuration and short runs of ventricular tachycardia. These ceased upon withdrawal of the catheter into the right auricle, but the episode was serious enough to cause signs of mild vascular collapse. One can easily visualize the possible development of ventricular fibrillation in the course of such an attack of ventricular tachycardia. This fatal complication has never been observed, as far as we know, by any of the groups investigating congenital heart disease. Death

\* Coronary sinus catheterization technique for studying coronary blood flow and myocardial metabolism in vivo. *Proc. Soc. Exper. Biol. and Med.* 66: 571, 1947.

is known to have occurred suddenly, however, in the case of two adult cardiacs, one of them with a long history of angina pectoris, but in neither instance was the catheterization performed with electrocardiographic control. It may be hypothesized that early recognition of ectopic beats or of runs of ventricular tachycardia would have led to withdrawal of the catheter, thus possibly preventing these accidents.

A paroxysm of auricular tachycardia has been reported by Johnson et al. in a case of interauricular septal defect. We have observed two such episodes in the course of our studies. In one case, in which previous attacks of a similar nature had been noted, the paroxysm ceased spontaneously after a short time, in the other it stopped immediately after displacement of the catheter.

**Blood loss:** The volume of blood withdrawn during the procedure is not less than 100 cc. This amount constitutes a fairly large proportion of the total circulating blood volume in young children and infants, the loss of which is likely to upset the steady state of the circulation. In such cases it is therefore important to estimate the volume of blood withdrawn during each step of the procedure and to replace it with an equivalent amount of donor blood.

The following figures illustrate the various locations of the tip of the catheter as seen on the fluoroscopic screen or recorded on x-ray films during the studies.

The first seven figures show the catheter with the tip located successively in (1) the right auricle, (2) the right ventricle pointing toward the apex, (3) the outflow tract of the right ventricle, (4) the truncus of the pulmonary artery near the valve and (5) near the bifurcation, (6) the left pulmonary artery, (7) the right pulmonary artery.



FIG. 1



FIG. 2

Figure 1: Catheter inserted in the right saphenous vein. The tip is located in the right auricle.

Figure 2: Catheter inserted in the right saphenous vein. The tip has been advanced through the tricuspid valve into the right ventricle and is pointing toward the apex.



FIG. 3

Figure 3: Catheter inserted in the left median basilic vein (Case 1). The tip has been advanced through the tricuspid valve into the right ventricle, pointing cephalad, and is located in the outflow tract. As premature ventricular contractions are apt to occur in this location, the catheter should never be left here without electrocardiographic control; and even with such control it should preferably be left for only a short time.



FIG 4



FIG 5

**Figure 4:** Catheter inserted in the left median basilic vein (Case 2) The tip is just on the arterial side of the pulmonary valve

**Figure 5:** Same case as Figure 4 The tip is near the bifurcation of the main pulmonary artery



FIG 6



FIG 7

**Figure 6:** Catheter inserted in the left median basilic vein (Case 13) The tip is in the left pulmonary artery

**Figure 7:** Catheter inserted in the right saphenous vein (Case 9). The tip is in the right pulmonary artery. In this position the tip of the catheter can often be seen moving back and forth several centimeters

Placement of the tip of the catheter in the various locations illustrated in the above seven figures is attempted routinely. It is usually easy unless tricuspid insufficiency or pulmonic atresia is present.

The remaining figures picture some of the unusual paths taken by the catheter in septal defects and complex cardiac anomalies.

In the next group of four figures the catheter tip has been introduced through an interauricular septal defect into the left auricle and then into one of the pulmonary veins or has been moved from the right auricle directly into a pulmonary vein.





FIG 8



FIG 9

**Figure 8:** Catheter inserted in the right saphenous vein (Case 9). The tip has been introduced through an interauricular septal defect into the left auricle. In comparing with Figure 2, where the catheter is passing through the tricuspid valve into the right ventricle, note that the curve here is higher and less acute

**Figure 9:** Catheter inserted in the right saphenous vein (Case 8). The tip, introduced into the left auricle through an interauricular septal defect, has been placed in a left pulmonary vein.



FIG. 10

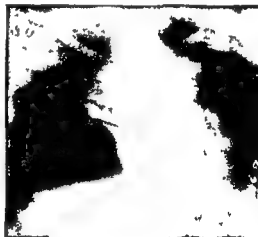


FIG 11

**Figure 10:** Catheter inserted in the right saphenous vein. The tip, introduced into the left auricle through an interauricular septal defect, has been placed in a left superior pulmonary vein. The exact location was ascertained by study of the pattern of the blood pressure tracing and by blood gas analysis. Otherwise, it could be mistaken for the left innominate vein or even a left pulmonary artery (compare with Fig. 6).

**Figure 11:** Catheter inserted in the right saphenous vein (Case 9). The tip has been introduced from the right auricle directly into a right pulmonary vein. Without the confirmation of the blood gas analysis, it could be assumed that the tip was in the right hemi-azygos vein.

In the following group of five pictures the catheter tip has been introduced into the left ventricle through an interventricular septal defect or into an overriding aorta.



FIG. 12



FIG. 13

**Figure 12:** Catheter inserted in the right saphenous vein (Case 12). The tip has been introduced through the tricuspid valve into the right ventricle and through an interventricular septal defect into the left ventricle. The location was confirmed by fluoroscopy in the oblique position (see Fig. 13), by recording the blood pressure (see Fig. 25), and by analysis of blood samples. With the catheter crossing the ventricular septum, the dangers of multiple premature contractions and ventricular tachycardia must constantly be kept in mind. To introduce the catheter into the left ventricle is not advisable.

**Figure 13:** The same case in the left anterior-oblique position.



FIG. 14



FIG. 15

**Figure 14:** Catheter inserted in the right median basilic vein. The tip has been introduced through the right ventricle into an overriding aorta with a left aortic arch. The position was confirmed by blood pressure tracings (see Fig. 27) and blood gas analysis.

**Figure 15:** The same case in the left anterior-oblique position.



FIG 16

**Figure 16:** Catheter inserted in the right median basilic vein (Case 16). The tip has been introduced through the right ventricle into an overriding aorta with a right aortic arch

The next four figures illustrate some of the unusual x-ray aspects observed in a case of dextrocardia (Case 17) with multiple congenital defects of the heart.



FIG. 17



FIG 18

**Figure 17:** Catheter inserted in a right median basilic vein. The tip was then introduced successively in a right superior vena cava, an auricular chamber with low blood pressure, and a left superior vena cava.

**Figure 18:** Catheter reinserted in the same right median basilic vein and seen to be crossing into a left superior vena cava. The tip was later introduced into a cardiac chamber with low blood pressure, then into a chamber with high blood pressure, the pattern of which was characteristic of the ventricle; and was finally placed in the pulmonary artery, identified only by the blood pressure tracings.



FIG 19



FIG 20

**Figure 19:** The tip of the catheter is in a cardiac chamber with low blood pressure (auricular pattern)

**Figure 20:** Here the tip is in a cardiac chamber with high blood pressure (ventricular pattern)

Determination of the location of the tip of the catheter is considerably helped by recording the blood pressure tracings and by immediately developing and examining the films. By watching the deflection of the manometer's light beam on the reflecting mirror of the recording apparatus, it is generally possible to ascertain, once the catheter is connected with the manometer, whether the tip of the catheter is in the right auricle, the right ventricle, or the pulmonary artery, as the patterns of the curves are quite distinct. The cyclic variations in the left auricle are of greater amplitude than those in the right auricle, and it requires a close inspection of the record to decide whether the tip of the catheter is in the left auricle or within the tricuspid valve area of the right ventricle. It is the timing of the large upward deflection, in relation to the P and QRS waves of the electrocardiogram, that provides the best means for differentiation. The blood pressure tracings respectively in the two ventricles and in the pulmonary artery and aorta are similar in form. They can only be differentiated by the height of pressures if normal conditions prevail in the right heart, as the systolic pressure in the left heart is about five times higher than in the right. Therefore, if the blood pressures are extremely elevated in the right heart, differentiation is based upon fluoroscopic observations and the results of the blood gas analysis.

The most characteristic patterns of blood pressures as recorded in the right auricle, the left auricle, the right ventricle, the left ventricle, the pulmonary artery, and the aorta are illustrated in Figures 21 to 26.

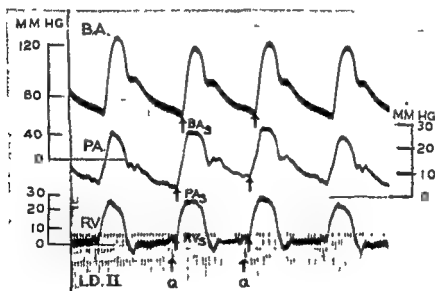


FIG. 21

Figure 21: Normal blood pressure patterns in the brachial artery, the pulmonary artery, and the right ventricle. In the pulmonary artery and right ventricular tracings the upward deflection starts after the beginning of the QRS complex. The slope of the downward deflection is very steep in the right ventricle, corresponding to the isometric relaxation preceding the plateau-like diastolic curve, and contrasts with the progressively downward diastolic curve in the pulmonary artery. As seen from the scale, the systolic pressure is less than 30 mm. Hg in the right ventricle and in the pulmonary artery.

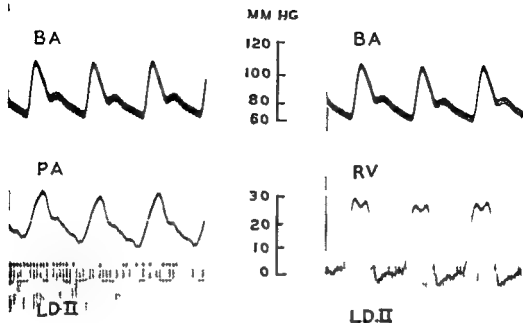


FIG 22

Figure 22: Blood pressure tracings in the brachial artery, the pulmonary artery, and the right ventricle in a case of interauricular septal defect. The blood pressure recordings were taken through a double lumen catheter without change in its position and in rapid succession. In spite of a large increase in pulmonary blood flow, the blood pressures in the pulmonary circulation were normal. This corresponds to similar observations made in normal subjects, indicating that a threefold increase in pulmonary blood flow does not cause any increase in pulmonary arterial pressure. The portion of the ascending slope corresponding to ejection in the right ventricle is much steeper than the analogous portion in the pulmonary artery. This difference in slope may be due in part to physical characteristics of the pulmonary arterial system (deformability of the large vessels, resistance in the small vessels, blood mass, etc.), in part to the velocity of ejection and probably in some part to the recording of pressure through the catheter.

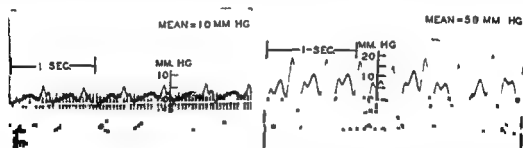


FIG 23

Figure 23: Blood pressure pattern in both auricles. Tracings taken in the right and left auricles of an infant of 5 months with an interauricular septal defect. To the left, the tracing taken in the right auricle: the main upward deflection (A) starts after the P wave of the electrocardiogram and before the QRS. It corresponds to the auricular systole. The downward slope of the A wave is interrupted by a plateau, followed by a dip, both corresponding to isometric contraction and beginning ejection in the right ventricle. The pressure variations during the entire cycle are small. The normal mean pressure varies within narrow ranges, with an upper limit of about 10 mm Hg.

mm. Hg. To the right, the tracing taken in the left auricle: the main upward deflection, starting after the P wave but before the QRS complex, corresponds to the auricular systole. The downward stroke of the A wave merges with the marked dip corresponding to the left ventricular isometric contraction and ejection. The pressure rises immediately following this dip during the remainder of the ventricular systole. It drops markedly again at the time of the opening of the mitral valve. The height of the deflection corresponding to the auricular systole is much greater in the left than in the right auricle. The mean pressure in the left auricle is from 2 to 4 mm. Hg higher than in the right auricle.



FIG. 24

Figure 24: Blood pressure pattern simulating left auricular tracings. Tracing taken with the tip of the catheter in the tricuspid valve area. The upward deflection corresponds to the ventricular systole. It starts after the beginning of the QRS complex. The whole curve is an artefact. (For comparison see Fig. 23, right curve )

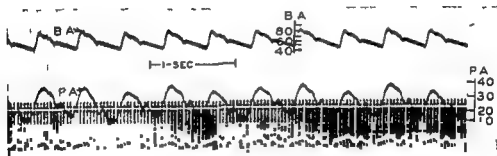


FIG. 25A

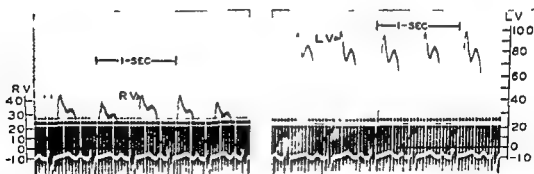


FIG. 25B

Figure 25: Blood pressure patterns in the left and right ventricles and in the pulmonary artery. Tracings taken in the case of a child of 3 years with an interventricular septal defect (Case 12). The left ventricular blood pressure tracing is identified by the large systolic deflection and the higher diastolic level. Otherwise, in form and in timing it is identical with that of the right ventricle.

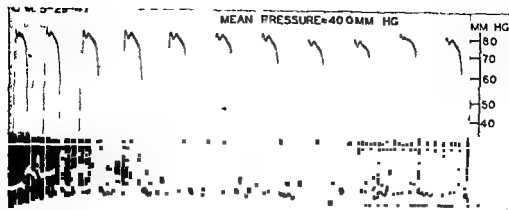


FIG 26

Figure 26: Blood pressure pattern in the right ventricle. Tracing taken in the case of a child of 6 years with an interauricular septal defect and right ventricular hypertension. It resembles in all respects a left ventricular tracing (see Fig 25). Right ventricular hypertension not associated with pulmonary stenosis, or overriding of the aorta, or transposition of the large vessels reflects either an increase in pulmonary vascular resistance, or hypertension in the left auricle and pulmonary veins, or a combination of both.

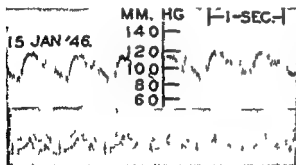
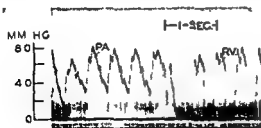


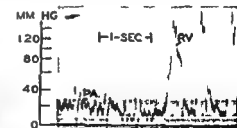
FIG 27

Figure 27: Blood pressure pattern in the aorta. Tracing taken in the case of a child of 12 years with a tetralogy of Fallot. The pattern is that of a central artery. Such tracings do not differ in form from those taken in the pulmonary artery. Therefore in instances of transposition of the large vessels analysis of the tracings cannot be of diagnostic value.

Aside from helping to determine the location of the tip of the catheter and to measure blood pressures in various cardiac cavities and large vessels, blood pressure recordings are of great value in the diagnosis of pulmonary stenosis. The next three figures show typical patterns of blood pressure tracings taken as the catheter is being withdrawn from the pulmonary artery into the right ventricle.



A WITHOUT PULMONARY STENOSIS



B WITH PULMONARY STENOSIS

Figure 28: Blood pressure tracings taken during withdrawal of the catheter from the pulmonary artery into the right ventricle (Case 10). To the left (A), typical finding in a subject without pulmonary stenosis but with pulmonary hypertension. The systolic pressures are identical in the pulmonary artery and the right ventricle. The slope of the downward curve and the dias-



tolic level are different. To the right (n), typical finding in a case of tetralogy of Fallot with pulmonary stenosis. The mean pressure in the pulmonary artery is very low. The pressure rises suddenly as the tip reaches the right ventricular outflow tract. Many vibrations are set up by the motion of the catheter during withdrawal.

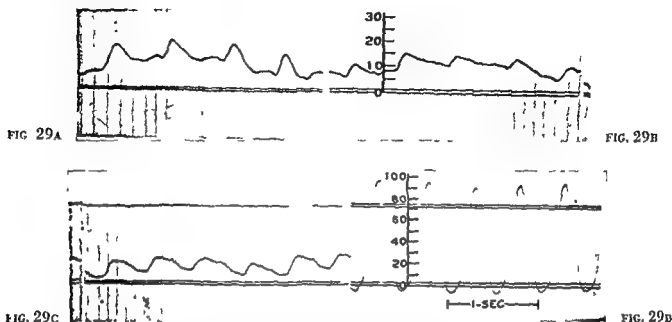


Figure 29: Blood pressure tracing taken during withdrawal of the catheter from the pulmonary artery into the right ventricle in a case of stenosis of the infundibulum.

The tracings were taken with the tip successively in the left branch of the pulmonary artery (A), the pulmonary trunk (B), the infundibular area of the right ventricle (C), and finally the right ventricle (D). The arterial pattern recognizable in the first two is less distinct in the infundibulum. When the tip reached a point in the ventricle very near the tricuspid valve, the pressure rose markedly and the blood pressure pattern became that of a right ventricle with hypertension. The exact location of the tip, as it is seen on the fluoroscopic screen when such a pressure change occurs, is an important landmark in differentiating between valvular and infundibular stenosis.

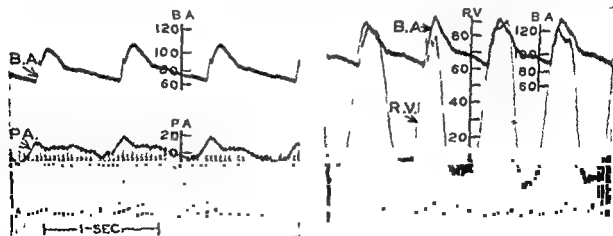
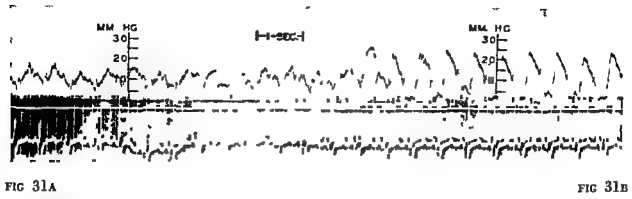


FIG. 30A

FIG. 30B

Figure 30: Blood pressure tracings taken with the catheter successively in the pulmonary artery and the right ventricle (Case 1). The same observations were made as in the previous case. The tracing in the infundibular region has not been reproduced.

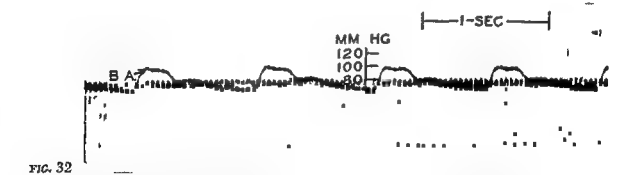
In the case of some patients with marked dilatation of the pulmonary artery of congenital origin, but without other malformations, a difference in systolic pressure exists between the pulmonary artery and the right ventricle, although the blood pressures in the latter may be within normal limits. A tracing taken in such a case is illustrated in the next figure.



**Figure 31:** Blood pressure tracings taken in a case of idiopathic dilatation of the pulmonary artery (Case 2)

The mean pressure in the pulmonary artery (A) is normal, the pulse pressure small. The systolic pressure in the right ventricle (B), although nearly 10 mm Hg higher than in the pulmonary artery, is normal. The same observation was made several times by moving the tip of the catheter back and forth in the valvular region. The abnormal expansibility of the dilated pulmonary artery, with its thin wall, is probably the chief cause of these pressure differences.

The blood pressure recordings from a peripheral artery may reveal abnormalities in the region of the aortic valve or further along the course of the aorta. Such a tracing is illustrated in the next figure.



**Figure 32:** Blood pressure tracing taken in a case of subaortic stenosis (Case 11). The plateau-like systole and the small pulse pressure are consistent with such a condition.

## Chapter IV

# FORMULAE FOR THE CALCULATION OF SYSTEMIC AND PULMONARY BLOOD FLOW AND OF BLOOD SHUNTS

## SYMBOLS

The data used for the calculation of the systemic or pulmonary blood flow and of various blood shunts are represented in the formulae by the following symbols

**Oxygen content in blood samples:** The letter "C" (for concentration) with a subscript indicating the location is used as a symbol to represent the oxygen content in the sample expressed in cubic centimeters per liter of blood

$C_{PA}$  = Oxygen concentration in the pulmonary artery

$C_{RV}$  = Oxygen concentration in the right ventricle

$C_{RA}$  = Oxygen concentration in the right auricle

In individuals without an abnormal communication between the left and the right heart, the concentration of oxygen in these three chambers or vessels usually checks within 3 cc per liter of blood. It differs exceptionally by as much as 6 cc per liter of blood.

$C_{SVC}$  = Oxygen concentration in the superior vena cava

$C_{IVC}$  = Oxygen concentration in the inferior vena cava

$C_{VC}$  = Oxygen concentration in the venae cavae, which by definition is derived

from the following formula  $\frac{C_{SVC} + C_{IVC}}{2}$

$C_{VC}$  is a calculated figure, as a rule not significantly different from  $C_{RA}$  in individuals without congenital heart defects. Occasionally the two columns of blood joining in the right auricle may remain separate as far as the right ventricle (streamline flow).

$C_{Ao}$  = Oxygen concentration in the aorta or in any peripheral artery

$C_{LV}$  = Oxygen concentration in the left ventricle

$C_{LA}$  = Oxygen concentration in the left auricle

$C_{PV}$  = Oxygen concentration in a pulmonary vein

Unless there is a shunt from right to left, the concentration of oxygen in the peripheral artery is identical with that in the aorta, the left ventricle, the left auricle, and the pulmonary veins. The normal oxyhemoglobin saturation in the arterial blood is 96 per cent or better, except in cases of interauricular septal defect, where  $C_{LA}$  may be measured by direct sampling; the assumption is made in the calculation of pulmonary blood flow that  $C_{LA}$  corresponds to a blood 96 per cent saturated with oxygen.

**Oxygen consumption (O<sub>2</sub>)** The oxygen consumption is expressed in cubic centimeters per minute. It is measured under basal metabolic conditions and in a steady state. The oxygen consumption in the tissues is equal to the oxygen intake in the lungs under these conditions.

**Systemic and pulmonary blood flow:** The systemic and the pulmonary blood flow are expressed in liters per minute and are represented by the following symbols

$Q_s$  = Systemic blood flow

$Q_{PA}$  = Blood flow in the pulmonary artery

$Q_{PC}$  = Pulmonary capillary blood flow

The pulmonary blood flow is equal to the pulmonary capillary blood flow in all instances, except when anastomoses to the pulmonary arterial system are supplied by the aortic system beyond the pulmonary artery or its main branches. In these instances  $Q_{PC}$  is greater than  $Q_{PA}$ .

**Blood shunts:** Blood shunts between the right heart and the pulmonary artery, and the left heart and the aorta, expressed in liters per minute are represented by the following symbols, which indicate the direction and the location of the shunt

- $\lambda$  = Right to left shunt of blood
- $Y$  = Left to right shunt of blood
- $Y_1$  = Shunt of blood from pulmonary veins or the left auricle into the right auricle
- $Y_2$  = Shunt of blood from the left into the right ventricle
- $Y_3$  = Shunt of blood from the aorta into the pulmonary artery
- $R$  = Blood flow in regurgitation through a cardiac valvular region, usually the pulmonic valve

### FORMULAE

The following formulae have been derived for the various calculations of systemic blood flow, pulmonary blood flow, and blood shunts

**Estimated systemic blood flow**

$$Q_s = \frac{O_2}{C_{Ao} - C_{RA} \text{ or } C_{VC}} \quad (1)$$

**Estimated pulmonary blood flow:** There are two cases to consider

*First case*  $Q_{PA} = Q_{PC}$ , then

$$Q_{PA} = \frac{O_2}{C_{LA} - C_{PA}} \quad (2)$$

This formula can only be used if no anastomoses are supplied by the aortic system to the pulmonary circulation beyond the point of sampling. It is essentially the same formula as formula 1 because oxygen consumption in the tissues is equal to oxygen intake by the lungs,  $C_{LA}$  is equal to  $C_{Ao}$  and  $C_{PA}$  is equal to  $C_{RA}$ .

*Second case*  $Q_{PA}$  is less than  $Q_{PC}$

When it is suspected that anastomoses are supplied by the aortic system to the pulmonary circulation beyond the point of collection of blood samples in the pulmonary artery ( $C_{PA}$ ), then  $Q_{PC}$  cannot be measured accurately, but it may be estimated as being between the two following limits

$$\frac{O_2}{C_{LA} - C_{PA}} \leq Q_{PC} \leq \frac{O_2}{C_{LA} - C_{Ao}} \quad (3)$$

In the left part of the formula it is assumed that the pulmonary capillary flow is supplied entirely by the pulmonary artery, in the right part, it is assumed that the capillary flow is supplied entirely by the aortic system. Obviously the pulmonary capillary blood flow lies somewhere between the limits thus defined. An estimate of this value may be clinically useful in cases where it is important to decide whether the pulmonary arterial blood flow is reduced.

**Blood shunts** There are several cases to consider, depending upon the oxygen concentration of the blood in various samples

*First case*  $C_{RA}$  is greater than  $C_{VC}$

Mixture in the right auricle of blood shunting from the left auricle or pulmonary vein with blood flowing from the venae cavae is suspected here. Another interpretation would be an interventricular septal defect with tricuspid insufficiency. Clinical examination and the special pattern of the blood pressure tracing in the right auricle would help in recognizing such a combination. The formula used to calculate the shunt is as follows

$$Y_1 = Q_s \left( \frac{C_{RA} - C_{VC}}{C_{LA} - C_{RA}} \right) \quad (4)$$

*Second case*  $C_{RV}$  is greater than  $C_{RA}$

Mixture in the right ventricle of left ventricular blood shunting through an interventricular septal defect with blood flowing from the right auricle is suspected. The difference may be present in all three of the samples from the right ventricle or only in the sample from the outflow tract. The formula used to calculate this shunt is

$$Y = Q_s \left( \frac{C_{RV} - C_{RA}}{C_{LV} - C_{RV}} \right) \quad (5)$$

If in addition  $C_{RA}$  is greater than  $C_{VC}$  the following formula is used

$$Y = (Q_s + Y_1) \left( \frac{C_{RV} - C_{RA}}{C_{LV} - C_{RV}} \right) \quad (6)$$

*Third case*  $C_{PA}$  is greater than  $C_{RV}$

In this case mixture in the pulmonary artery of blood shunting from the aorta through a patent ductus arteriosus or an aortico pulmonary septal defect with blood flowing from the right ventricle is suspected. Large differences in the various blood specimens sampled in the truncus and the main branches of the pulmonary artery depend upon (a) the location of the tip of the catheter with regard to the column of blood coming from the aorta and (b) the degree of mixing which takes place. The choice of a figure to use in the calculation may cause variation in the estimated blood shunts over a considerable range. The formula used to calculate this shunt is

$$Y_2 = Q_s \left( \frac{C_{PA} - C_{RV}}{C_{AO} - C_{PA}} \right) \quad (7)$$

*Fourth case*  $C_{PA}$  greater than  $C_{RV}$  and  $C_{RV}$  greater than  $C_{RA}$

There is a choice between two formulae when it is found that in addition to an aortico pulmonary shunt  $C_{RV}$  is greater than  $C_{RA}$ . The choice depends on whether the clinical findings support the diagnosis of (a) an aortico pulmonary shunt associated with an interventricular septal defect or (b) an aortico pulmonary shunt associated with pulmonic incompetence.

In the first instance the interventricular shunt is calculated from formula 5 and the aortico pulmonary shunt from the following formula

$$Y_2 = (Q_s + Y) \left( \frac{C_{PA} - C_{RV}}{C_{AO} - C_{PA}} \right) \quad (8)$$

In the second instance the aortico pulmonary shunt is calculated from formula 7 and the volume of pulmonic regurgitation is calculated from the following formula

$$R = Q_s \left( \frac{C_{RV} - C_{RA}}{C_{PA} - C_{RV}} \right) \quad (9)$$

If an interventricular septal defect is associated with a patent ductus arteriosus and pulmonic incompetence, it is impossible to measure separately the flow of blood shunting from the left to the right ventricle and regurgitating through the incompetent valve

It should be noted that, in the absence of an aortico pulmonary shunt, the diagnosis of pulmonic regurgitation cannot be established on the basis of blood gas analyses

*Fifth case* The oxyhemoglobin saturation of the arterial blood is reduced below the normal value of 96 per cent

A right to left shunt is suspected, and the blood flow through the shunt is calculated from the following formula

$$X = Q_s \left( \frac{C_{LA} - C_{AO}}{C_{LA} - C_{VC}} \right) \quad (10)$$

This shunt most commonly occurs between the two auricles or between the right ventricle and an overriding aorta. Very rarely it may take place between the pulmonary artery and the aorta as a result of partial reversal of blood flow through a patent ductus arteriosus. In this instance for mula 10 is not applicable

In the case of an interventricular septal defect combined with overriding of the aorta in which there is a double blood shunt from left to right and from right to left, formulae 5 and 10 measure respectively (a) the volume of blood of left auricular composition which flows into the pulmonary artery and (b) the volume of blood of right auricular composition which flows into the aorta. The approximate volume of the columns of blood circulating in each direction, across the defect and becoming intermixed in a manner impossible to predict, may be calculated if equation 5 is expressed as follows

$$Y' = Q_s \frac{(C_{PA} - C_{RA})}{(C_{LA} - C_{PA})} \frac{(C_{AO} - C_{RA})}{(C_{LA} - C_{RA})} \quad (5a)$$

$$\text{and if } X = Q_s + Y' - Q_{PA} \quad (11)$$

In some instances a special technique is required to differentiate the oxygen unsaturation of the arterial blood, produced by a shunt of mixed venous blood from that due to inadequate oxygenation of capillary blood in the lungs. This technique consists in collecting arterial blood samples while the subject is breathing pure oxygen. If the shunt is 25 per cent or more, the arterial blood fails to become fully saturated with oxygen



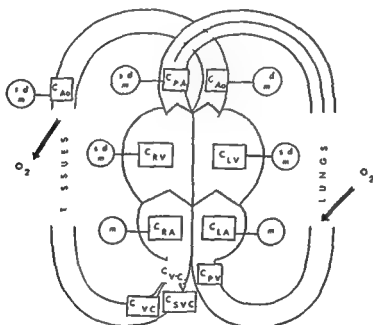
## **PART TWO**



## ILLUSTRATIVE CASES

A uniform plan has been followed in the presentation of the seventeen individual cases selected for discussion. The data and comments are given in the following order:


- 1 The clinical data, with emphasis on the cardiac signs and the x ray findings
- 2 The clinical diagnosis, prior to the physiological studies
- 3 The physiological data, presented in a schematic form (see the symbols listed on pages 26-27, the diagram illustrated below, and the key on the opposite page)
- 4 The calculations of blood flow and blood shunts computed on the basis of the oxygen content in the venous blood samples and of the oxygen consumption
- 5 Comments concerning the validity and significance of the observations made in the course of the physiological studies
- 6 The follow up history, including a brief description of operative or autopsy findings when available
- 7 A general comment combining and correlating all the observations




SCHEME OF THE DIAGRAMS

# KEY TO THE DIAGRAMS



## Blood Oxygen Content

	cc./lit. of blood == observed
	cc./lit. of blood == assumed or derived








## Blood Pressures in mm Hg

	s == systolic
	d == diastolic
	m == mean

## Valvular Lesions

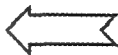
	Dilatation Competent valve Stenosis
	Incompetent valve Stenosis

## Blood Oxygen Saturation

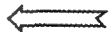
	0 to 20%
	21 to 40%
	41 to 60%
	61 to 70%
	71 to 80%
	81 to 90%
	90% and over

## Shunts—Direction and Volume

Shunt equal to or greater than systemic flow



Shunt between 33% and 100% of systemic flow

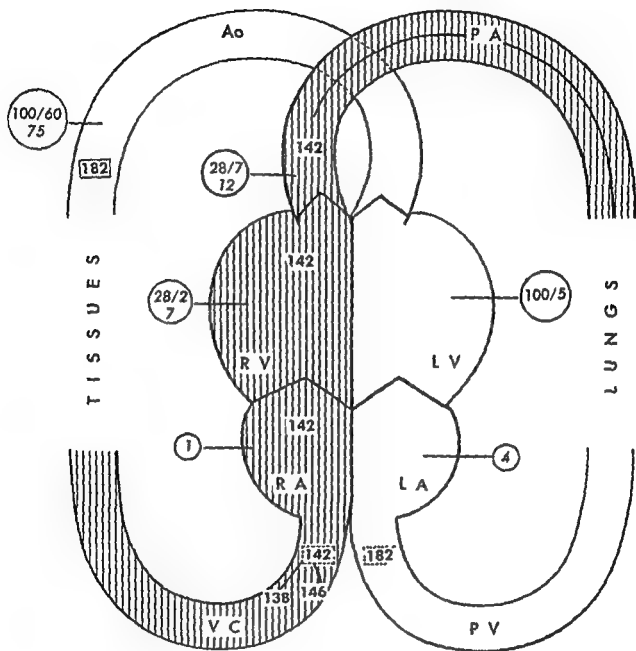


Shunt less than 33% of systemic flow



# APPROXIMATE HEMODYNAMIC DATA

IN A NORMAL CHILD—AGE 3 YEARS, WEIGHT 14 KG.



Arterial blood oxygen capacity . . = 192 cc/lit.

Arterial blood oxygen saturation = 96%

Oxygen consumption . . . at rest = 100 cc/min.

## ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) . . . . . 2.50 lit/min.

Pulmonary ( $Q_{rA}$ ) . . . 2.50 lit/min.

# Case 1

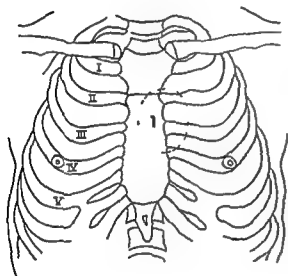
## INFUNDIBULAR PULMONARY STENOSIS POST-STENOTIC DILATATION OF PULMONARY ARTERY

**Clinical data:** Age 11 years Weight 25.4 Kg (56 lb-) F

**History:** This child was somewhat slender but otherwise well developed. No clubbing, dyspnea, or easy fatigue had been noted.

**Physical examination:** There was no cyanosis or dyspnea on considerable exercise.

### HEART



**Size:** Slightly enlarged to left

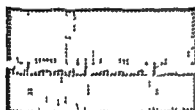
**Thrill:** Coarse systolic close to sternum at II left interspace

**Sounds:** P<sub>2</sub> faint

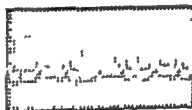
**Murmurs:** 1. Long, loud, rather coarse systolic over upper left precordium, maximum at III left interspace at sternal edge, did not transmit well toward left clavicle

**Arterial blood pressure:** 112/72 mm Hg, after exercise, 118/76 mm Hg

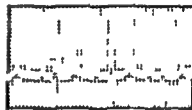
### ELECTROCARDIOGRAM



LEAD I

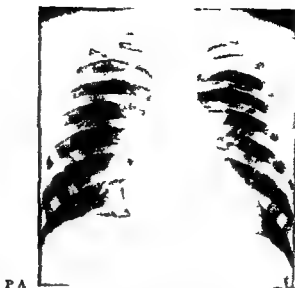


LEAD II



LEAD III

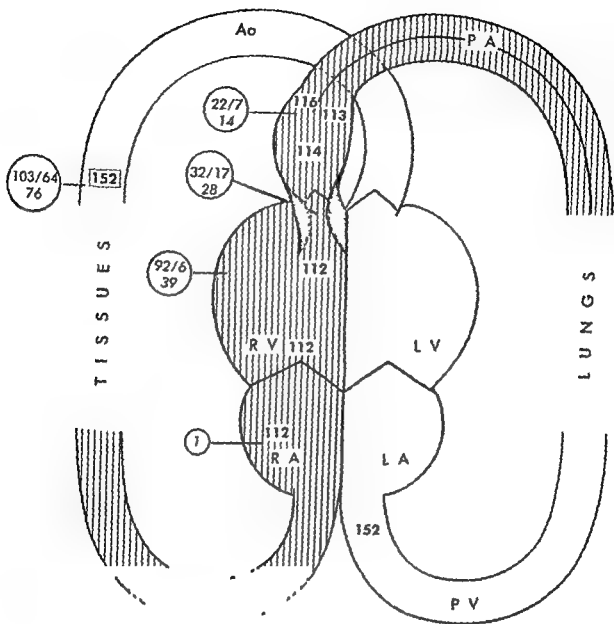
Slight right axis deviation. Normal conduction time. Slight notching of P waves.



There was slight transverse enlargement of the heart with the apex lifted well above the diaphragm. In the region of the pulmonary area there was a prominent rounded shadow which showed very little pulsation. A slight concavity was present immediately below this shadow. In the right anterior oblique view there was some flattening in the region of the outflow area of the right ventricle with prominence of the pulmonary artery above. In the left anterior oblique view the right ventricle showed moderate enlargement but the left ventricle did not appear enlarged. There was no increase in the vascular markings in the lung fields.

## CLINICAL DIAGNOSIS

*Pulmonary stenosis with post stenotic dilatation of the pulmonary artery.* The presence of other anomalies which are frequently associated with pulmonary stenosis, such as an interventricular septal defect and overriding of the aorta in various degrees, was considered unlikely on the basis of the physical signs and the size and shape of the heart as shown by x ray, and in view of the lack of any suggestion of cyanosis after considerable exercise.



Arterial blood oxygen capacity  $\approx 162 \text{ cc/lit}$   
 Arterial blood oxygen saturation  $\text{at rest} \approx 95\%$   
 Arterial blood oxygen saturation following mild exercise  $\approx 94\%$   
 Oxygen consumption  $\text{at rest} \approx 186 \text{ cc/min}$

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ )  $1.78 \text{ lit/min}$   
 Pulmonary ( $Q_{pA}$ )  $1.78 \text{ lit/min}$

## COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration by catheter showed the right ventricle to be enlarged and the main pulmonary artery to be considerably dilated. Since the blood samples from the right auricle and the right ventricle were identical in oxygen content the presence of an interventricular septal defect was ruled out. While the catheter was being withdrawn from the pulmonary truncus to the right ventricle, the abrupt change in pressure occurred low within the midsternal shadow (see Fig 30). This indicated that the stenosis was presumably at the infundibular portion of the right ventricle rather than at the pulmonary valve. There was a marked degree of systolic hypertension in the right ventricle. To explain the fall in pressure between the infundibular region and the left branch of the pulmonary artery the presence of turbulent flow in the dilated post stenotic pulmonary truncus was suggested.

## GENERAL COMMENTS

The physiological study confirmed the clinical diagnosis and ruled out the presence of other congenital defects. The remarkable clinical tolerance of the marked degree of systolic hypertension in the right ventricle should be emphasized.

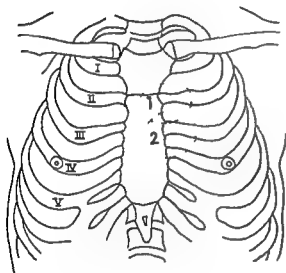
## Case 2 || IDIOPATHIC DILATATION OF PULMONARY ARTERY

**Clinical data:** Age 6 years Weight 22.7 Kg (50 lbs) F

**History:** Development was normal. The child's exercise tolerance was never decreased from normal. A murmur was noted shortly after birth.

**Physical examination:** There was a moderately deep funnel depression at the lower sternum. No dyspnea or cyanosis was noted on exertion.

### HEART



**Size** Increased at base to left

**Thrill** Moderate systolic along II left inter space

**Sounds** P only slightly accentuated

**Murmurs** (See stethogram)

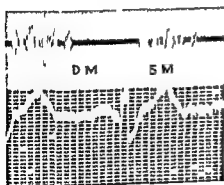
- 1 Loud, very long, coarse systolic, maximum at II left interspace
- 2 Less loud, blowing, decrescendo diastolic, louder in III than II left interspace

**Arterial blood pressure** 105/70 mm Hg, after exercise, 112/75 mm Hg

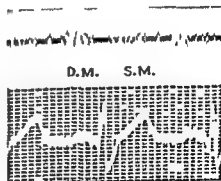
### ELECTROCARDIOGRAM

Slight right axis deviation. Right bundle branch block, type B (see Lead II, below)

### STETHOGRAM



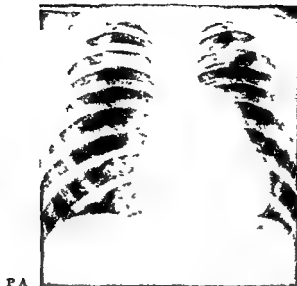
II LEFT INTERSPACE



III LEFT INTERSPACE



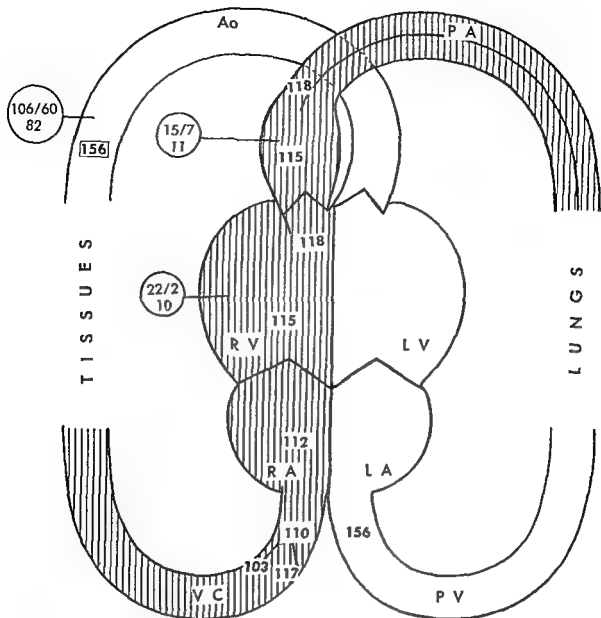
## X-RAY AND FLUOROSCOPY



There was striking enlargement, though little increase in pulsation, of the main pulmonary artery and its left branch; the other pulmonary vessels were not enlarged or pulsating abnormally. The left ventricle and the auricles did not seem to be enlarged; the right ventricle was slightly enlarged.

### CLINICAL DIAGNOSIS

*Idiopathic dilatation of the pulmonary artery, truncus and left branch, with pulmonic incompetence.* This diagnosis, as against that of a patent ductus arteriosus, was based on the following points: (1) the systolic murmur was of equal intensity throughout the phase and lacked intensification at the end of systole, which is frequently noted in cases of a patent ductus; (2) the diastolic murmur was not only entirely different in quality, as it was much more blowing and softer than the systolic, but also differed in location, the point of maximum intensity being below that of the systolic murmur and transmitted well down the left sternal border; (3) the second pulmonic sound was not significantly increased in intensity; (4) the pulse pressure was not wide and the diastolic blood pressure did not drop after exercise; (5) x ray and fluoroscopic findings of a very large but only moderately pulsating main and left pulmonary artery, without enlargement and increased pulsations in the rest of the pulmonary arterial tree.



Arterial blood oxygen capacity — 162 cc/lit  
 Arterial blood oxygen saturation at rest — 98%  
 Oxygen consumption at rest = 127 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 3 10 lit/min  
 Pulmonary ( $Q_{rA}$ ) 3 10 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the main pulmonary artery and its left branch with the tip of the catheter clearly demonstrated a marked degree of dilatation (see Fig 31). The oxygen content of blood samples from the pulmonary artery and the chambers of the right side of the heart showed no significant difference, indicating that there was no shunt of oxygenated blood at any point. Thorough mixing of the venae cavae blood columns was still not complete in the right ventricle. The pressure tracings showed a small difference in systolic pressure between the pulmonary artery and the right ventricle. This may be chiefly explained by an increase in the expansibility of the pulmonary arteries due to the thinness and dilatation of their walls. The role played by turbulent flow is probably less important. In the absence of any degree of hypertension in the right ventricle, organic pulmonary stenosis of even mild degree was considered unlikely.

## GENERAL COMMENTS

The physiological study revealed no evidence of a patent ductus arteriosus and supported the most favored clinical diagnosis of idiopathic dilatation of the pulmonary artery. Without angiographic studies it was not possible to decide whether hypoplasia of the ascending aorta was associated with this malformation. In the absence of a shunt of highly oxygenated blood above the pulmonary valve the diagnosis of pulmonic incompetence based on the clinical finding of a diastolic murmur could not be confirmed.

### Case 3

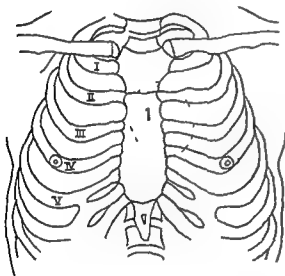
## PATENT DUCTUS ARTERIOSUS MILD HYPERTENSION IN PULMONARY CIRCULATION

**Clinical data:** Age 3 years Weight 13.6 Kg (30 lbs.) F

**History:** The child had always been very active, without evident dyspnea or cyanosis. A murmur was first reported at 1½ years.

**Physical examination:** Her development was normal for her age. There was slight pallor.

### HEART



**Size** Slightly enlarged

**Thrill** Systolic, maximum at II and III left interspaces

**Sounds** P. masked by murmurs

**Murmurs** I Very loud, continuous machinery type systolic and diastolic murmurs, maximum at II and III left interspaces, transmitted moderately to right and downward along left side of sternum. No murmurs at apex.

**Arterial blood pressure** 94/48 mm Hg, after exercise, 98/40 mm Hg.

### ELECTROCARDIOGRAM

**Normal** No deviation of electrical axis

## X-RAY AND FLUOROSCOPY

P A



L A-O



R A-O



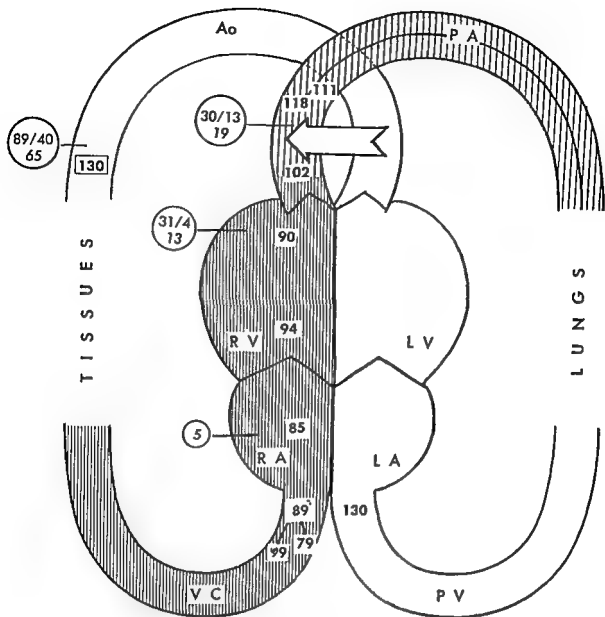
R A-O



The heart showed a slight general increase in size. There was considerable prominence of the left ventricle, especially in the left anterior-oblique view, and of the pulmonary artery segment in the postero-anterior and right anterior-oblique views. Slight increase in size of the left auricle was seen in the esophagram. The right auricle was not enlarged. There was an increase in the pulmonary vascular markings in the hilar areas, but no notably expansile pulsations of these vessels were present.

### CLINICAL DIAGNOSIS

*Patent ductus arteriosus* with little enlargement of the heart. This diagnosis was made on the basis of classical types of murmurs, x-ray findings, and wide pulse pressure. In view of the relatively slight increase in pulmonary markings, the amount of shunt through the ductus was thought not to be large.



Arterial blood oxygen capacity — 140 cc/lit  
 Arterial blood oxygen saturation at rest = 92%  
 Oxygen consumption (estimated) at rest = 95 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 2.38 lit/min  
 Pulmonary ( $Q_{pA}$ ) 4.98 lit/min

#### ESTIMATED BLOOD SHUNT

Aortico pulmonary ( $\gamma_s$ ) 2.60 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

The data on oxygen content in the right ventricle and right auricle suggested that there was in complete mixing in these chambers of the two very different streams of venous blood returning through the superior and inferior venae cavae. The marked increase in oxygen content in the truncus and main branches of the pulmonary artery indicated a shunt of oxygenated blood from the aorta. Because of its higher oxygen content the blood sample from the right pulmonary artery was probably more representative of blood shunting from the aorta than of truly mixed pulmonary arterial blood. The estimated pulmonary blood flow given in the table was based on the oxygen content of the sample in the left pulmonary artery. If calculated from the sample from the right pulmonary artery it would be 7.91 lit/min. There was only a very slight increase in the mean and diastolic pressures in the pulmonary artery.

## OPERATIVE FINDINGS

At operation (by Dr. Herbert C. Muer) a rather short patent ductus about 1.5 cm. in diameter was found and ligated with subsequent complete disappearance of the murmurs and thrill.

## FOLLOW UP

Over the next 9 months there was a decrease in the prominence of the pulmonary segment and the left ventricular shadow and in the accentuation of the pulmonary vascular markings.

## GENERAL COMMENTS

This is a typical case of patent ductus arteriosus without complications such as pulmonary hypertension and pulmonic incompetence. The physiological study confirmed the clinical diagnosis and suggested that the size of the communication and the amount of blood shunting from the aorta were relatively small. These findings are consistent with the clinical observations of only a moderate degree of cardiac enlargement and moderate increase of the pulmonary vascular shadows without expansile pulsation. It should be noted that with the relatively small increase in the pulmonary flow returning to the left ventricle there was no diastolic murmur heard at the apex.

With the pressures in the pulmonary artery almost normal it appears obvious that vascular changes in the pulmonary arterioles and/or hypertension in the somewhat dilated left auricle were minimal.

## PATENT DUCTUS ARTERIOSUS

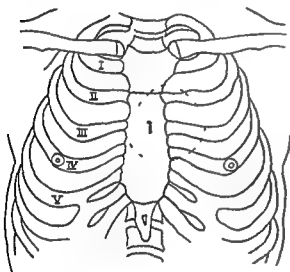
## MODERATE HYPERTENSION IN PULMONARY CIRCULATION

**Clinical data:** Age 3 years Weight 12.7 Kg (28 lb.) F

**History.** There had been a trace of cyanosis, noted during two attacks of bronchopneumonia. Exercise tolerance was somewhat decreased.

**Physical examination.** The child was slightly underdeveloped. There was no clubbing or evidence of cyanosis, but mild dyspnea appeared on fairly active play. The breath sounds were increased in the posterior left chest behind the heart.

## HEART



**Size** Enlarged

**Thrills** Coarse, systolic at II and III left interspaces

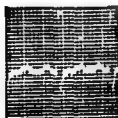
**Sounds** P. increased

**Murmurs** (See stethiogram, page 48)

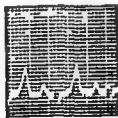
- I Continuous, coarse, "train in tunnel" type, most intense at end of systole and early diastole. Heard widely over upper and mid precordium, maximum at II and III left interspaces. No apical murmurs.

**Arterial blood pressure** 95/15 mm Hg, after exercise, 98/35 mm Hg

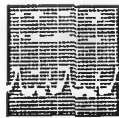
## ELECTROCARDIOGRAM



LEAD I



LEAD II

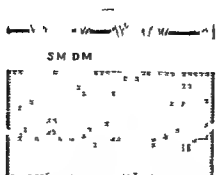


LEAD III

No axis deviation or conduction defect.



## STETHOGRAM



II LEFT INTERSPACE

## X RAY AND FLUOROSCOPY

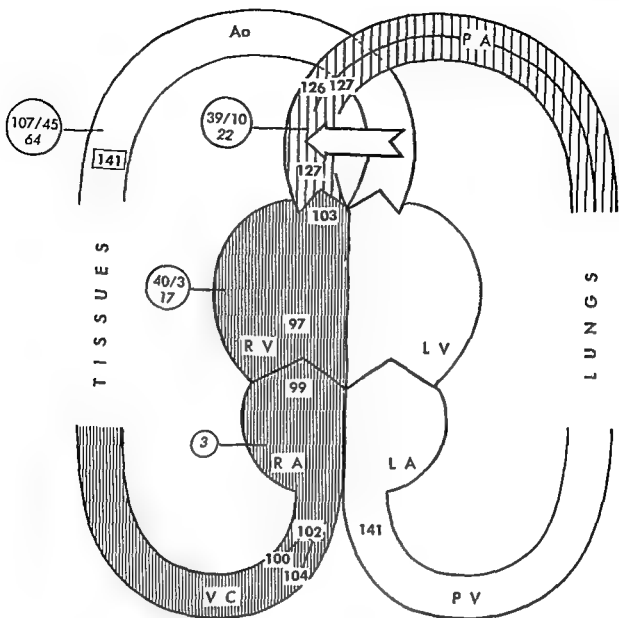


P A

There was general enlargement of the heart, and the apex was low. The outflow tract of the right ventricle and the pulmonary artery were enlarged as seen in both the postero anterior and the right anterior oblique view. In the left anterior oblique view the left ventricle was quite strikingly enlarged posteriorly. There was moderate increase in the size of the right auricle, and the barium filled esophagus showed compression from an enlarged left auricle. The lung fields showed considerably increased pulmonary vascular markings, especially in the hilar areas with moderately expansile pulsations.

## CLINICAL DIAGNOSIS

*Patent ductus arteriosus* with considerable increase in pulmonary flow and enlargement of the heart. This case exhibited the typical clinical signs: (1) classical murmurs, (2) wide pulse pressure, (3) drop in diastolic blood pressure after exercise, and (4) hilar dance and increased pulmonary vascular markings.



Arterial blood oxygen capacity 148 lit/min  
 Arterial blood oxygen saturation at rest 97%  
 Oxygen consumption at rest - 95 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 2.37 lit/min  
 Pulmonary ( $Q_{PA}$ ) 6.78 lit/min

#### ESTIMATED BLOOD SHUNT

Aortico-pulmonary ( $Y_s$ ) 4.11 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

The presence of an aortico pulmonary shunt was established from the physiological study, with no evidence of other shunts. The blood samples from the main pulmonary artery and both branches were identical in oxygen content, suggesting that the blood was very well mixed. The calculation of the pulmonary circulation is therefore reasonably accurate. There was no evidence of pulmonic regurgitation of any significance. Only a moderate degree of hypertension was present in the pulmonary circulation. This was of interest in view of the fairly large calculated blood shunt from the aorta into the pulmonary artery and of a total pulmonary blood flow about three times that of the systemic blood flow.

## OPERATIVE FINDINGS

At operation (by Dr. Charles W. Lester) a ductus about 1 cm. long and 0.7 cm. in diameter was found. The pulmonary artery and the outflow tract of the right ventricle were seen to be markedly dilated. The ductus was ligated, and the murmurs and thrill disappeared.

## GENERAL COMMENTS

The physiological study confirmed the clinical diagnosis. It is surprising that in this case, where the calculation showed a fairly large pulmonary blood flow and where enlargement of the left auricle and ventricle was noted, there was no diastolic murmur at the apex indicating "relative," non organic mitral stenosis.

An increase in pressure in the dilated left auricle was presumed to be the cause of the moderate increase in pressure in the pulmonary artery.

It should be emphasized that in the presence of classical clinical signs of a patent ductus arteriosus catheterization of the heart is not necessary as a preoperative diagnostic procedure, unless there is reason to suspect an associated anomaly, particularly pulmonary stenosis.

## Case 5

# PATENT DUCTUS ARTERIOSUS INTERVENTRICULAR SEPTAL DEFECT MARKED PULMONARY HYPERTENSION

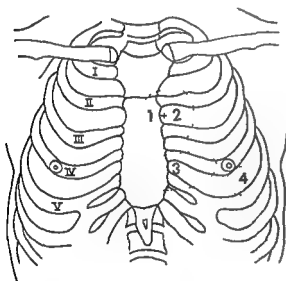
**Clinical data** Age 8 years Weight 18.4 Kg (40½ lbs) F

**History** The child had had two attacks of pneumonia and frequent bronchitis. She was somewhat late in walking and had always been less active than other children, showing easy fatigue and dyspnea on climbing more than one flight of stairs. There was no history suggestive of rheumatic fever.

**Physical examination** Fragile underdeveloped child. There was no cyanosis but definite dyspnea appeared with moderate exertion.

## HEART

**Inspection** Slight precordial bulge to left of sternum.



**Size** Enlarged downward and out to left.

**Thrill** Moderate systolic maximum at III left interspace.

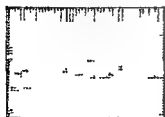
**Sounds** First at apex increased P markedly accentuated.

**Murmurs**

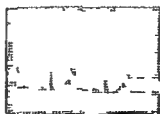
- 1 Long loud coarse systolic
- 2 Less coarse early to mid diastolic both maximum at II left interspace 1 and 2 became continuous after exercise
- 3 Systolic over lower left sternal area transmitted to left higher pitched and less coarse than 1 over pulmonic area
- 4 Mid diastolic localized at apex different timbre and lower pitch than 2 over pulmonic area

**Arterial blood pressure** 125/76 mm Hg  
after exercise 125/48 mm Hg

## ELECTROCARDIOGRAM



LEAD I



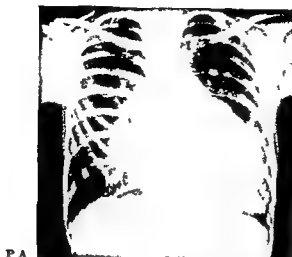
LEAD II



LEAD III

Slight notching of P<sub>1</sub> and P<sub>2</sub> diphasic QRS complexes of increased amplitude in Leads I and III  
No conduction defect or deviation of the electrical axis

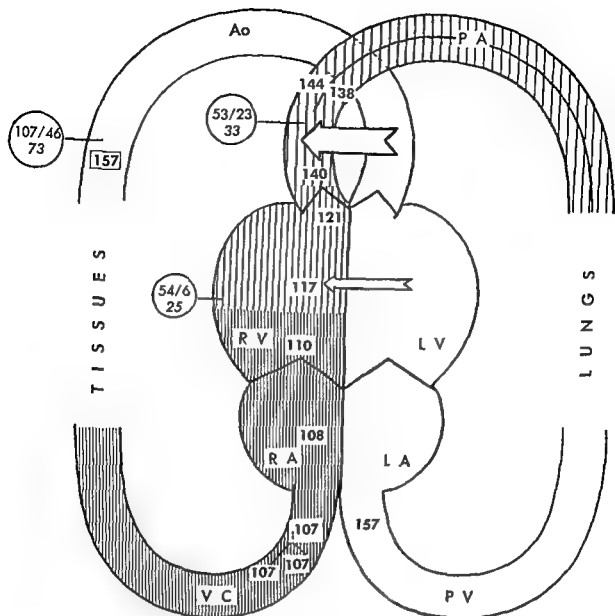
## X-RAY AND FLUOROSCOPY



X-ray and fluoroscopic examination showed generalized enlargement of the heart. The left ventricle was enlarged outward and down in the postero-anterior view and markedly overrode the spine in the left anterior-oblique view. Enlargement of the right ventricle was observed in all positions. The pulmonary artery segment was prominent, and there was congestion of the smaller pulmonary vessels as well as a moderate degree of expansile pulsation noted in the vessels in the hilar region. Both auricles were definitely enlarged.

### CLINICAL DIAGNOSIS

The clinical diagnosis was (1) a patent ductus arteriosus on the basis of murmurs and thrill over the pulmonic area and blood pressure findings; (2) an interventricular septal defect based on the finding of a separate systolic murmur over the lower precordium. The presence of the second shunt was also suggested by the striking enlargement of the heart, especially the left ventricle. The separate diastolic murmur at the apex was felt to be due to a "relative" mitral stenosis caused by enlargement of the left ventricle and very large pulmonary return flow.



Arterial blood oxygen capacity 173 cc/lit.  
 Arterial blood oxygen saturation at rest = 92%  
 Oxygen consumption at rest 152 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 3.10 lit/min  
 Pulmonary ( $Q_{PA}$ ) 9.50 lit/min

#### ESTIMATED BLOOD SHUNT

Left to right ventricle ( $\chi_2$ ) 0.90 lit/min  
 Aorta to pulmonary artery ( $\chi_1$ ) 5.50 lit/min

#### COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the right auricle and ventricle by catheter showed both to be very large. A run of auricular tachycardia developed during exploration of the enlarged auricle but subsided after the tip of the catheter was moved.

The presence of an arterial shunt from the aorta into the pulmonary artery was demonstrated with a calculated total pulmonary flow of 9.50 lit/min, about three times that of the systemic

flow. The difference in oxygen content between the samples taken in the right auricle and the out-flow area of the right ventricle could indicate either a shunt of blood from the left to the right ventricle through an interventricular septal defect or pulmonic regurgitation. There was a marked degree of systolic hypertension in the right ventricle and the pulmonary artery.

### OPERATIVE FINDINGS AND FOLLOW-UP



At operation (by Dr. Herbert C. Maier) the pulmonary artery was found to be considerably enlarged, with a thrill present. The patent ductus arteriosus, measuring approximately 1 cm. in width and 1 cm. in length, was ligated with prompt cessation of the thrill.

After operation the murmurs over the pulmonic area disappeared. As was expected, in view of the interventricular defect, the murmur over the lower precordium persisted with the original intensity and quality. The apical mid-diastolic murmur became very short and faint.

During a 4 months' follow-up period there was a marked gain in weight as well as improvement in cardiac function capacity. The reduction of the vascular markings and the decrease in size of all the cardiac cavities were striking. The child's condition was obviously greatly improved by the elimination of one of the two blood shunts.

### GENERAL COMMENTS

This case was of interest on several accounts. (1) In view of the higher oxygenation of the right ventricular blood compared with the right auricular blood in a patient with a patent ductus arteriosus, the presence of a second congenital defect (namely, an interventricular defect) rather than of pulmonic regurgitation, was postulated on the basis of a separate systolic murmur over the lower precordium. For comparison see Case 7, where the diagnosis of pulmonic regurgitation was made on the basis of similar findings in the oxygen content of the blood but different physical signs. The persistence of the systolic murmur over the lower precordium after ligation of the patent ductus in the present case confirmed the original interpretation. (2) The marked decrease in intensity of the mid-diastolic murmur at the apex suggests that after reduction in the pulmonary blood flow, following ligation of the patent ductus, the degree of "relative" mitral stenosis was reduced. (3) The ligation of the patent ductus was followed by decided improvement in the child's general status as well as in her cardiac functional capacity, even in the presence of an interventricular shunt.

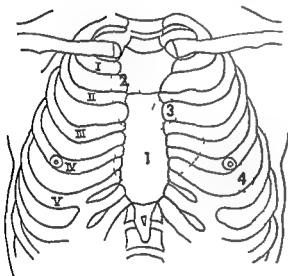
The marked elevation of pressures in the pulmonary artery was probably related both to pulmonary vascular changes and to hypertension in the dilated left auricle.

**Clinical data:** Age 4 years Weight 10 Kg (22 lbs) F

**History:** The mother had German measles during the sixth week of pregnancy. The child had had annual attacks of bronchopneumonia during which only slight cyanosis was present. There was moderate dyspnea on exertion.

**Physical examination:** There was marked underdevelopment, with muscular hypotonia, bilateral cataracts, and pigeon breast with deep indentations occurring bilaterally at the level of the diaphragm.

## HEART



Arterial blood pressure 73/35 mm Hg

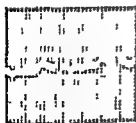
**Size** Enlarged to left and downward

**Thrills** 1 Fine systolic at iv left interspace  
2 Coarser systolic at suprasternal notch and into neck

**Sounds** First at apex very loud A<sub>2</sub> present, equal to P<sub>2</sub>.

**Murmurs** 1 Very loud, harsh systolic over lower precordium, maximum at iv left interspace  
2 Coarser systolic, maximum at ii right interspace, transmitted to neck and axilla  
3 Very faint, short diastolic at ii left interspace  
4 Short mid diastolic at apex when supine

## ELECTROCARDIOGRAM



LEAD I



LEAD II

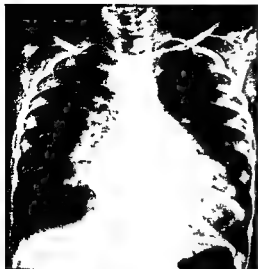


LEAD III

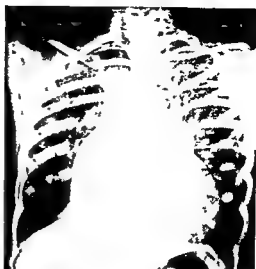
Normal conduction, high T waves and deep Q waves in Lead II and III. No significant axis deviation.



## X-RAY AND FLUOROSCOPY



P-A



L A-O



R A-O

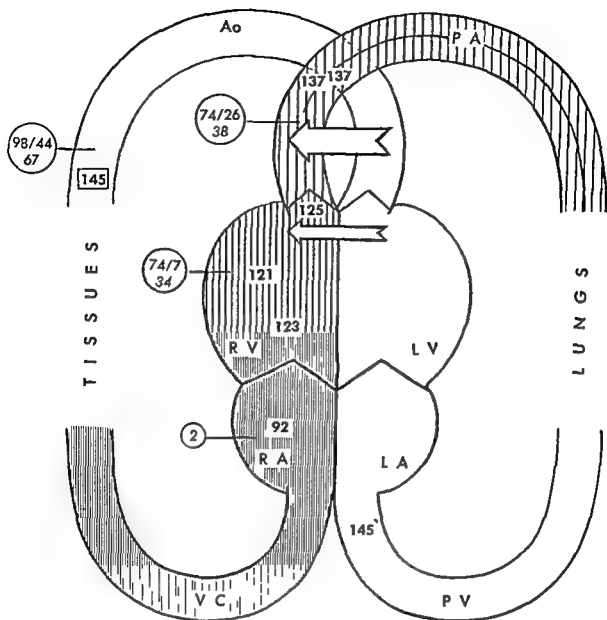


RT. LAT.

There was marked enlargement of the left ventricle in the postero-anterior view and posteriorly in the left anterior-oblique view. The aortic shadow appeared small. The right ventricle and the pulmonary arc were enlarged; increased and collapsing pulsations of the pulmonary vessels were seen. The region of the right auricle was prominent. Note the displacement by chest deformity in the right lateral view.

### CLINICAL DIAGNOSIS

Multiple cardiac anomalies: (1) an *interventricular septal defect* as the cause of the systolic murmur over the lower precordium, (2) *subaortic stenosis* causing the systolic murmur described over the right upper precordium, and (3) a *possible patent ductus arteriosus* causing the diastolic murmur at the second left interspace and the wide pulse pressure and adding to the expansile pulsations in the pulmonary vascular shadows. The enlarged size of the right ventricle and the pulmonary vessels was considered due to the increased volume flow resulting from left-to-right shunts through the interventricular septal defect and possibly through a patent ductus. The marked increase in blood return from the lungs would account in part for the enlargement of the left auricle and ventricle, and explain the mid-diastolic murmur at the apex on the basis of a "relative" mitral stenosis. In such a case, with shunting of a large proportion of the circulation into the right heart, the small volume to the systemic circulation would account for the poor general physical development, the low arterial blood pressure, and the small aortic shadow.



Arterial blood oxygen capacity = 153 cc/lit.  
 Arterial blood oxygen saturation at rest = 96%  
 Oxygen consumption at rest = 108 cc/min

#### ESTIMATED BLOOD FLOW

#### ESTIMATED BLOOD SHUNT

Systemic ( $Q_s$ )      ■ 04 lit/min  
 Pulmonary ( $Q_{pA}$ )    ■ 13 50 lit/min

Left ventricle to right ventricle ( $Y_2$ )    ■ 86 lit/min  
 Aorta to pulmonary artery ( $Y_1$ )        ■ 60 lit/min.

#### COMMENTS ON THE PHYSIOLOGICAL STUDY

The study demonstrated a large blood shunt from the aorta into the pulmonary artery. However, it was considered that the pulmonary flow as calculated might be too high, the blood samples from both branches of the pulmonary artery being more representative of the stream flowing from the aorta to the pulmonary artery than of a complete mixture of blood flowing from the right ventricle and the aorta. A marked degree of hypertension was present in the pulmonary circuit. In addition

the marked increase in oxygen content of the right ventricle blood as compared with the right auricle blood was assumed to be due to a fairly large volume of blood shunted from the left to the right ventricle through an interventricular defect. Pressure tracings taken in the brachial artery failed to reveal the plateau like type of curve characteristic of stenosis in the aortic area

### OPERATIVE AND AUTOPSY FINDINGS

At operation (by Dr Charles W Lester) the base of the heart was found to be lower than normal, with the apex rotated up in such a way that the arch of the aorta lay almost behind the heart. There was a palpable thrill in the aorta, the subclavian artery, and the mediastinum over the ductus. During exploration in the region of the ductus the heart stopped suddenly, and attempts at resuscitation failed.

At autopsy (courtesy of Dr W W Brandes, Roosevelt Hospital) the chief findings were (1) a high interventricular septal defect 13 mm in diameter, (2) a patent ductus arteriosus, 5 mm in diameter, 13 mm long (3) marked hypertrophy of the left ventricle, moderate hypertrophy of the right ventricle, (4) thickening of the aortic valve cusps, (5) considerable narrowing of the lumen of the pulmonary precapillaries.



VIEW OF THE OPEN LEFT VENTRICLE SHOWING THE HYPERTROPHIED MUSCLE AND THE SEPTAL DEFECT IMMEDIATELY UNDER THE FIBROSED AORTIC VALVES

### GENERAL COMMENTS

The physiological study in this case demonstrated the presence of a patent ductus arteriosus which had been uncertain on clinical grounds. This was confirmed at operation and autopsy. The diagnosis of an interventricular septal defect was made both on clinical grounds and from catheterization studies, and was also confirmed at autopsy. The fibrosis of the cusps of the aortic valve found at autopsy may explain the site of maximum intensity and direction of transmission of the thrill and systolic murmur at the base, although it is probable that the patent ductus arteriosus contributed to these signs. Pulmonary vascular changes were indicated by marked elevation of pressures in the pulmonary artery.

# Case 7

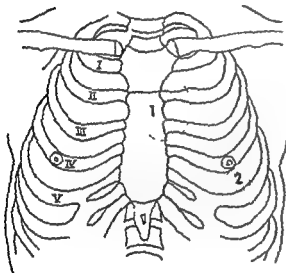
## PATENT DUCTUS ARTERIOSUS AND MARKED PULMONARY HYPERTENSION (STUDIED BEFORE AND AFTER LIGATION)

**Clinical data:** Age 3 years Weight 11.8 Kg (26 lbs) F

**History:** The child had had severe bronchopneumonia at 2½ years, with cyanosis and moderate congestive failure. While under continued observation she gradually developed a diastolic murmur over the pulmonic area, a mid diastolic murmur at the apex, enlargement of the heart, and increase in pulmonary vascular markings.

**Physical examination:** Development was normal. Tachycardia and moderate dyspnea were present with exercise.

### HEART



**Size:** Enlarged to left.

**Thrill:** Slight systolic at 11 left inter-space.

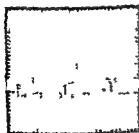
**Sounds:** P<sub>2</sub> increased.

**Murmurs:** 1 Rough continuous, machinery type over pulmonic area.

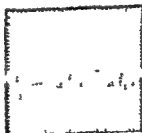
2 Short mid diastolic at apex, best heard when supine in left lateral position.

**Arterial blood pressure:** 108/60 mm Hg, after exercise, 112/50 mm Hg.

### ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Normal conduction time. No axis deviation.

## X-RAY AND FLUOROSCOPY

P A



L A-O



Enlargement of right ventricle and pulmonary arc. Pulmonary vascular markings increased, with hilar dance. Left ventricle increased in size, especially posteriorly. Right auricle moderately enlarged. Left auricle slightly compressed the barium-filled esophagus.

### CLINICAL DIAGNOSIS

*Patent ductus arteriosus* with enlargement of heart. Diastolic murmur at apex interpreted as due to "relative" mitral stenosis caused by large pulmonary flow returning to enlarged left heart.

### OPERATIVE FINDINGS

At operation (by Dr. Charles W. Lester) a ductus measuring approximately 1 cm. in length and 1 cm. in width was found. On ligation of this vessel the thrill previously palpable along the pulmonary artery ceased and the heart rate decreased.

### FOLLOW-UP

Five weeks post-operatively: Complete disappearance of murmurs over pulmonic and apical areas. Fluoroscopy and x-rays showed some decrease in the size of the pulmonary artery, which, however, was still quite prominent. Right ventricle only slightly decreased.

Five months post-operatively: Interval course very satisfactory, with 5 pounds gain in weight. Pulse and respiratory rate normal; returned to base level quickly after vigorous exercise.  $P_2$  no longer accentuated. No murmurs. Blood pressure: 96/60 mm. Hg; after exercise, 100/60 mm. Hg

P A

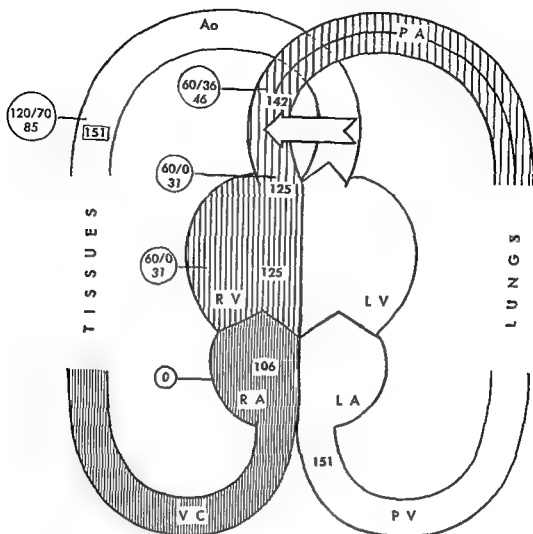


L A-O



X-rays showed a further decrease in the size of the heart and pulmonary artery and normal pulmonary vascular shadows.

# PRE OPERATIVE CATHETERIZATION



Arterial blood oxygen capacity — 162 cc/lit  
 Arterial blood oxygen saturation at rest = 95%  
 Oxygen consumption at rest = 92 cc/min

## ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 2.01 lit/min  
 Pulmonary ( $Q_{PA}$ ) 10.20 lit/min

## ESTIMATED BLOOD SHUNT

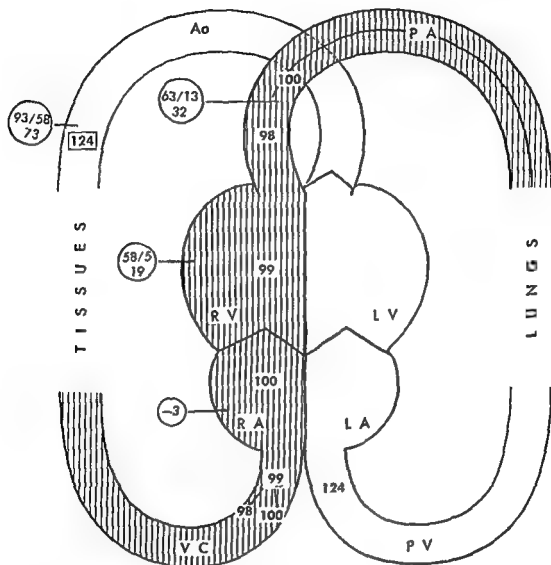
Aortico pulmonary ( $Y_a$ ) 8.16 lit/min  
 Pulmonic regurgitation (R) 2.8 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

**Before operation** The oxygen content in the blood samples demonstrated the presence of an arterial shunt into the pulmonary artery. The calculated volume of blood shunt was very large and the total pulmonary flow was five times that of the systemic flow. There was marked hypertension, both systolic and diastolic in the pulmonary artery. The difference in oxygen content between the right atrium and the right ventricle indicated a flow of highly oxygenated blood into the right ventricle. The interpretation could be either an associated interventricular septal

defect or pulmonic incompetence with regurgitation of highly oxygenated pulmonary arterial blood through a dilated pulmonic ring. In the absence of a separate systolic murmur below the pulmonic area the diagnosis of pulmonic incompetence was postulated.

### POST OPERATIVE CATHETERIZATION—5 WEEKS



Arterial blood oxygen capacity = 133 cc/lit  
 Arterial blood oxygen saturation at rest = 91%  
 Oxygen consumption at rest = 92 cc/min

### ESTIMATED BLOOD FLOW

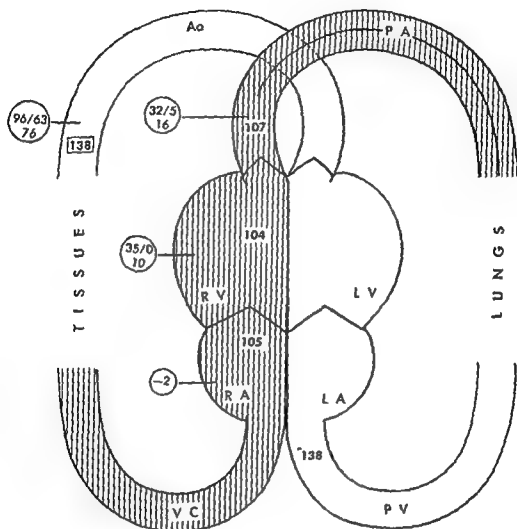
Systemic ( $Q_s$ ) 3.83 lit/min  
 Pulmonary ( $Q_{PA}$ ) 3.83 lit/min

### COMMENTS ON THE PHYSIOLOGICAL STUDY

Five weeks after operation. There was no longer any evidence of shunt into either the pulmonary artery or the right ventricle. This confirmed the earlier impression that the evidence of

a shunt into the right ventricle found at pre operative catheterization had been due to pulmonic incompetence. The systolic pressure of the pulmonary artery was still markedly elevated, while the diastolic pressure had reached a normal level. The pulse pressure was therefore very large.

### POST OPERATIVE CATHETERIZATION—5 MONTHS



Arterial blood oxygen capacity 144 cc/lit  
 Arterial blood oxygen saturation at rest = 98%  
 Oxygen consumption at rest 99 cc/min

### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 3.00 lit/min  
 Pulmonary ( $Q_p$ ) 3.00 lit/min

### COMMENTS ON THE PHYSIOLOGICAL STUDY

Five months after operation: The pressure in the right ventricle and the pulmonary artery had decreased strikingly and had almost returned to normal.



Clinical signs of a patent ductus arteriosus may be less clear cut in infants and young children than in older persons. This case presented an opportunity of observing over the period of a year the progressive appearance of the diastolic element and the machinery type murmur as well as a systolic thrill over the pulmonic area. During this period there was also clinical evidence suggestive of cardiac insufficiency and of an increasing volume of blood flow through the ductus. This was indicated by x ray findings of both relative and absolute increase in the size of the heart and pulmonary artery, and by increasing vascular markings in the lung fields with hilar pulsations of a more collapsing type. The development of a mid diastolic murmur limited to the apex in a child who showed no trace of rheumatic infection was interpreted as an added sign of increased return flow from the lungs to the left side of the heart with dilatation of both chambers and swirling of the blood as it passed through an organically normal mitral valve into the enlarged left ventricle.

The physiological studies in this case were of interest in showing a very large increase in pulmonary blood flow prior to ligation of the patent ductus arteriosus. In this respect the interpretation of the mid diastolic murmur at the apex as being due to increased return flow to the left heart was probably correct as it disappeared after ligation of the ductus when the pulmonary flow was proved to have returned to an almost normal figure.

It must be emphasized that in cases of arterial shunt into the pulmonary artery a higher oxygen content in the right ventricle than in the right auricle may be interpreted as the result either of an associated ventricular septal defect or of pulmonic incompetence. The latter diagnosis is suggested if there is evidence of considerable increase in pulmonary blood flow with considerable dilatation of the pulmonary artery. The difficulty of separating the systolic murmur of a high ventricular septal defect from the systolic component of a concomitant patent ductus arteriosus sometimes makes an exact interpretation of blood gas analysis impossible.

The presence of a high systolic pressure in the pulmonary artery 5 weeks after ligation of the patent ductus arteriosus at the same level as pre-operatively can only be explained by the presence of pulmonary vascular changes due to the very large pulmonary blood flow or to the persistence of left auricular dilatation with hypertension. The return to a normal blood pressure in the pulmonary artery after 5 months is evidence that the vascular changes or the left auricular dilatation or both were slowly reversible. Furthermore the large pulse pressure observed in the pulmonary artery at the time of the second study was presumptive evidence that pulmonic incompetence was still present. It is to be noted that this characteristic finding of valvular incompetence became apparent only after the patent ductus had been ligated and the communication with the aorta during diastole had been eliminated. The very large pulse pressure could be alternately explained by the continuance of the high pulmonary vascular resistance.

# Case 8

## INTERAURICULAR SEPTAL DEFECT MILD PULMONARY STENOSIS SUBACUTE BACTERIAL ENDOCARDITIS

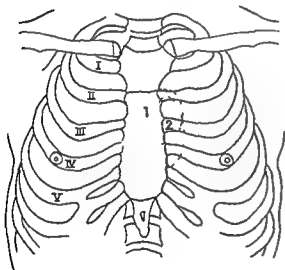
**Clinical data** Age 5 years Weight 13.6 Kg (30 lbs) F

**History** The child's physical development had been slow. Easy fatigability had always been noted.

**Present illness** Progressive weight loss, pallor and fever for 5 weeks. Condition critical on admission. Marked pallor, dyspnea and orthopnea present, tachycardia, gallop rhythm, heart markedly dilated, liver and spleen greatly enlarged. Many carious teeth. Positive blood cultures for *Streptococcus viridans* (600 colonies per cc.) confirmed the diagnosis of subacute bacterial endocarditis. Congestive failure gradually decreased on digitalis and repeated small transfusions. Clinical response to penicillin for 4 weeks was poor, although the organism was sensitive in vitro. It was therefore most important to decide whether a patent ductus arteriosus was present and if so to ligate it.

**Physical examination** No clubbing or cyanosis. Clinical signs of mild failure still present.

### HEART



**Size** Enlarged to right and left

**Thrill** Slight systolic over pulmonic area

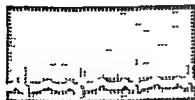
**Sounds** P slightly increased

**Murmurs** 1 Long loud harsh systolic at II and III left interspaces

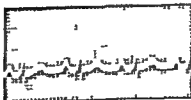
2 Soft blowing diastolic at II and IV left interspaces

**Arterial blood pressure** 80/55 mm Hg

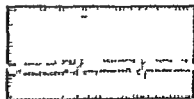
### ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

P waves prominent in Lead II. No deviation of electrical axis.

## X-RAY AND FLUOROSCOPY



P A

ON ADMISSION



P A

L A O

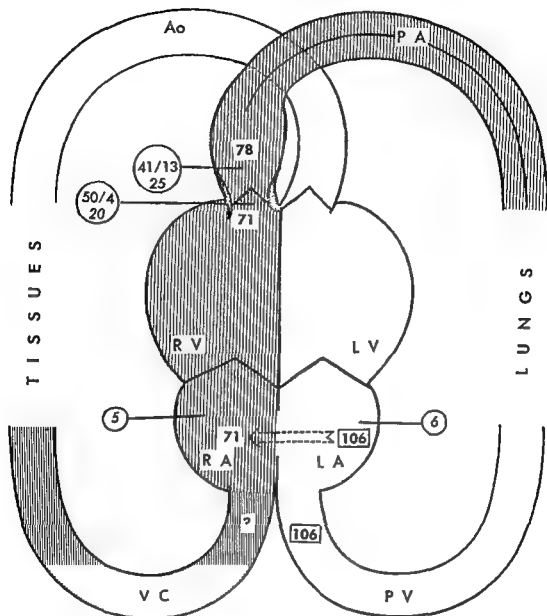
FOUR WEEKS LATER, AT TIME OF FIRST CATHETERIZATION

**Time of first catheterization:** Generalized enlargement of heart. Right auricle and ventricle increased in postero-anterior and left anterior-oblique views; pulmonary artery and branches very prominent with active collapsing type of pulsations noted. Left ventricle enlarged in postero-anterior and right anterior-oblique views. Left auricle compressed the barium-filled esophagus.

### CLINICAL DIAGNOSIS

The diagnosis of an *interauricular septal defect with pulmonic regurgitation* was more favored than that of a patent ductus arteriosus, on the basis of the following: (1) the degree of cardiac enlargement, especially of the right auricle, which persisted when other signs of congestive failure were receding; (2) the normal pulse pressure; and (3) particularly the soft, blowing character of the diastolic murmur transmitted down the left sternal border from the pulmonary area. The possibility that the clinical signs could be caused either by an interauricular septal defect with pulmonic incompetence or by a patent ductus arteriosus, especially in the presence of moderate congestive failure, was fully recognized and stressed the need for establishing the diagnosis more accurately.

# FIRST CATHETERIZATION



Arterial blood oxygen capacity = 112 cc/lit  
 Arterial blood oxygen saturation at rest = 96%  
 Oxygen consumption at rest = 102 cc/min

## ESTIMATED BLOOD FLOW

Pulmonary ( $Q_{RA}$ ) 3.00 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

First catheterization: The presence of an interauricular septal defect was established by passing the tip of the catheter from the right auricle through the left auricle into a left pulmonary vein. As blood samples from the superior and inferior venae cavae were not taken, the degree of

shunt from the left auricle to the right auricle could not be estimated nor the systemic blood flow calculated. A sample of blood taken from the brachial artery while the child was crying showed an oxygen saturation of 89 per cent, a significant reduction when compared to the oxygen saturation in the left auricular sample obtained while the child was quiet. This would indicate a mild degree of right-to-left shunt during physical exertion.

The pressure recordings in the right ventricle indicated a significant degree of hypertension. The systolic blood pressure in the pulmonary artery was elevated but somewhat lower than that in the right ventricle.

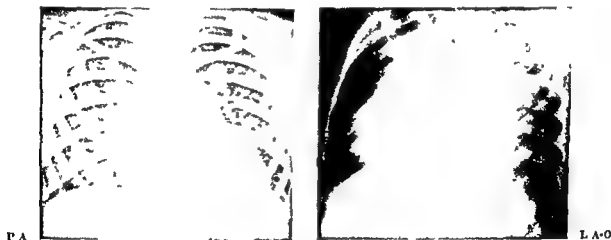
The diagnosis from the physiological study was an interauricular septal defect with probably a mild degree of pulmonary stenosis. The high pressure in the pulmonary artery was considered related to increased vascular resistance in the pulmonary circuit due to congestive failure.

### INTERVAL COURSE

**Up to time of second catheterization:** Penicillin was increased to 1 million units daily for 6 weeks with sterilization of the blood and continued clinical improvement up to the time of the second catheterization, done 4 months later.

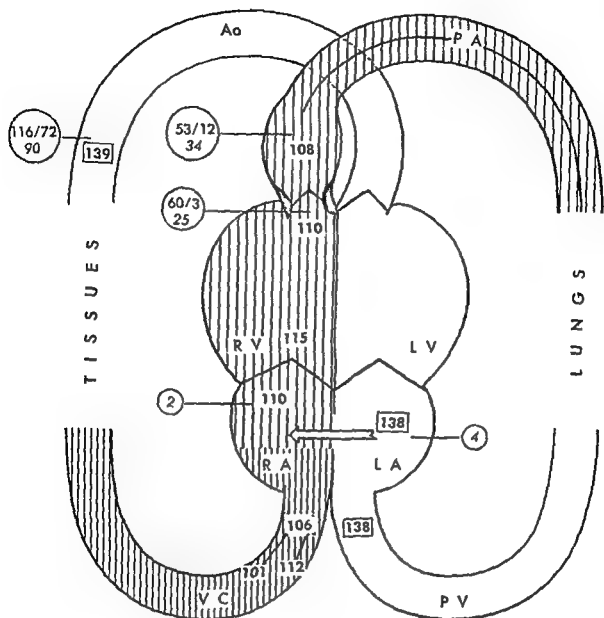
#### At time of second catheterization:

**Clinical data:** No signs of congestive failure or active infection. Decrease in size of heart. Thrill over pulmonic area more pronounced. Systolic murmur louder; diastolic murmur shorter and fainter.



**X-ray and fluoroscopy:** General decrease in size of heart, less congestion in lung fields. Collapsing type of pulsations of the main and hilar pulmonary vessels more striking.

## SECOND CATHETERIZATION



Arterial blood oxygen capacity      = 140 cc/lit.  
 Arterial blood oxygen saturation at rest = 98%  
 Oxygen consumption                      at rest = 102 cc/min

### ESTIMATED BLOOD FLOW

### ESTIMATED BLOOD SHUNT

Systemic ( $Q_s$ )                      8.09 lit/min  
 Pulmonary ( $Q_{PA}$ )                3.40 lit/min

Left to right auricle ( $Y_1$ )      0.31 lit/min

### COMMENTS ON THE PHYSIOLOGICAL STUDY

**Second catheterization:** The tip of the catheter was again directed into the left auricle and left pulmonary vein. The sampling of blood in the left auricle, right auricle, superior and inferior

venae cavae made possible the calculation of systemic blood flow and volume of blood shunted from the left to the right auricle. This shunt was shown to be very small.

A difference in systolic pressure between the pulmonary artery and the right ventricle was again demonstrated on repeated tracings, and the previous finding of pulmonary stenosis of a mild degree was therefore confirmed. There was still a marked degree of hypertension in the pulmonary circuit after clinical signs of failure had cleared. This now strongly suggested that the cause of the increased pulmonary resistance was due to permanent changes in the pulmonary vascular tree.

When compared with that of the previous study, the pulmonary blood flow showed an increase. Also, the mean right auricular blood pressure was significantly lower, which was to be expected as the clinical signs of decompensation were no longer present.

### GENERAL COMMENTS

By accurately establishing the diagnosis of an interauricular septal defect and eliminating that of a patent ductus, the use of cardiac catheterization prevented an unnecessary exploratory thoracotomy. In view of the extreme rarity of an interauricular septal defect as the site of subacute bacterial endocarditis, the finding of pulmonary stenosis on separate occasions, even though in very mild degree, threw significant light on this case, since a stenotic area is a more likely site of infection. With no shunt of highly oxygenated blood beyond the pulmonary valve, it was impossible to confirm the diagnosis of pulmonic incompetence from the data obtained by catheterization. This diagnosis therefore had to be based entirely on the finding of a characteristic murmur.

The persistence of a high systolic pressure in the pulmonary artery several months after recovery from cardiac failure can be adequately explained only on the basis of pulmonary vascular changes of a permanent nature. In this instance the presumed changes were not related to a large pulmonary blood flow. It is suggested that they may have been secondary to the subacute bacterial endocarditis and the prolonged episode of cardiac failure associated with it. It should be noted that these permanent vascular changes would adequately explain a rise in the systolic pressure in the pulmonary artery as the cardiac output increased following compensation.

In general, the passage of a catheter in the presence of subacute bacterial endocarditis is not recommended. In this case the danger of dislodging a fragment of vegetation by the catheter in the course of the diagnostic procedure was considered less of a hazard than subjecting the child to an exploratory thoracotomy.

# Case 9

## INTERAURICULAR SEPTAL DEFECT

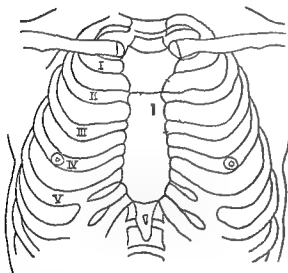
## ANOMALOUS IMPLANTATION OF A PULMONARY VEIN PULMONARY STENOSIS

**Clinical data:** Age 2½ years Weight 7.5 Kg (16½ lbs) F

**History:** This child was so markedly retarded that she never held up her head. She had had one attack of bronchopneumonia. No cyanosis had been observed.

**Physical examination:** Mongoloid features and hypotonia. Dentition was greatly delayed and defective. The palate was cleft.

### HEART



**Size:** Moderately enlarged to left

**Thrill:** Systolic in I and II left interspaces

**Sounds:** P<sub>2</sub> present although not noticeably accentuated

**Murmurs:** 1. Loud rather coarse systolic, maximum at II left interspace, not well transmitted toward left clavicle

**Arterial blood pressure:** 74/50 mm Hg

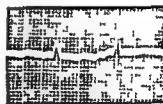
### ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Right axis deviation. No conduction defect.



## X-RAY AND FLUOROSCOPY



P.A.

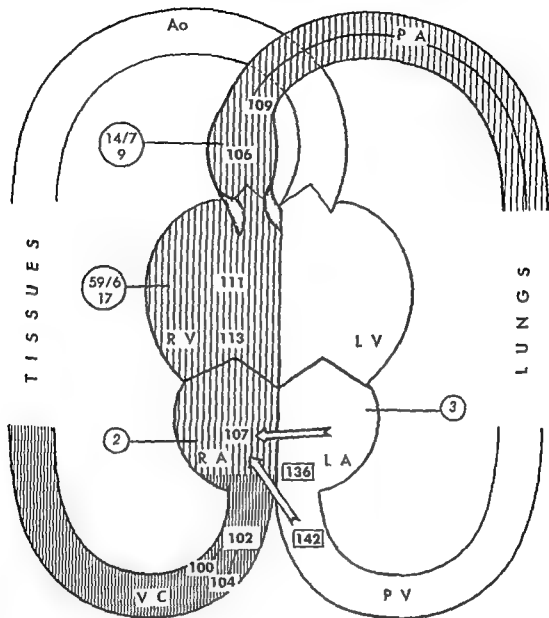


RT. LAT.

The right ventricle was enlarged, and there was fullness of the pulmonary arc but without striking pulsations in the latter area. The right auricle was moderately enlarged in both the postero-anterior and the right anterior-oblique view. No increase in vascular markings or congestion of the lung fields was noted.

### CLINICAL DIAGNOSIS

*Interauricular septal defect.* This diagnosis was based on the location of the murmur and the size of the right auricle, right ventricle, and pulmonary arc as seen in the x-ray.



Arterial blood oxygen capacity 147 cc/lit  
 Arterial blood oxygen saturation at rest 94%  
 Oxygen consumption (estimated) at rest — 59 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_A$ ) 1.17 lit/min  
 Pulmonary ( $Q_A$ ) 1.73 lit/min

#### ESTIMATED BLOOD SHUNT

Pulmonary vein and left auricle to right auricle ( $Y_1$ ) 0.26 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

The presence of an interauricular septal defect was demonstrated by placement of the catheter in the left auricle and a left pulmonary vein. Direct implantation of a right pulmonary vein in the right auricle was also demonstrated (see Figs. 11 and 11).

The amount of blood shunting from the left to the right side was small, and the calculated pulmonary blood flow was therefore not much increased. The differences in the oxygen content of the various samples from the right cardiac chambers and pulmonary artery were slightly above the upper limit of normal variation. This suggests the persistence of streamline flow as far as the pulmonary artery. In this case the presence of an interauricular septal defect could not have been diagnosed by comparing the oxygen content in both venae cavae and in the right auricle, as there was almost no difference.

Continuous pressure recordings demonstrated stenosis in the infundibular region of the right ventricle and marked elevation of blood pressure in the right ventricle.

## GENERAL COMMENTS

Catheterization studies, in addition to confirming the clinical diagnosis of an interauricular septal defect, demonstrated the anomalous implantation of a pulmonary vein and the presence of infundibular pulmonary stenosis which had not been suspected clinically. In retrospect it seems that pulmonary stenosis might have been suggested by the fact that there was no increase in the pulsations of the pulmonary arc and in the vascular markings of the lungs. Post-stenotic dilatation might explain the enlarged pulmonary artery. Furthermore, the demonstration of a fairly small volume of blood shunting into the right auricle and of only a relatively small increase in pulmonary blood flow might well account for the absence of pulsation and of pulmonary congestion. It should be noted that the small shunt into the right auricle was associated with but moderate enlargement of this chamber. In this case of interauricular septal defect with small pulmonary blood flow the finding of pulmonary stenosis explains the degree of enlargement of the right ventricle.

# IMPLANTATION OF PULMONARY VEINS IN RIGHT AURICLE OR INTERAURICULAR SEPTAL DEFECT

**Clinical data** Age 8 months Weight 4.8 Kg (10½ lbs) F

**History** This child was markedly retarded (Mongolian idiot). Before her birth her mother had had six other pregnancies; the next sibling was eight years older. There had been questionable cyanosis on rare occasions during long crying spells.

**Physical examination** There were typical Mongoloid facies and hypotonia. The liver edge was felt two fingerbreadths below the costal margin.

## HEART

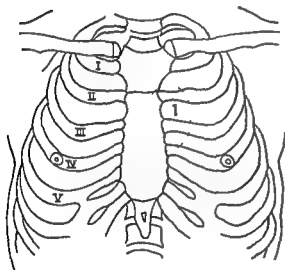
*Size* Enlarged to left and right.

*Thrills* None felt.

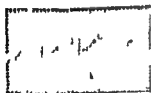
*Sounds* First sound at apex markedly increased. P<sub>2</sub> moderately increased.

*Murmurs* 1. Long loud systolic over left precordium; maximum at II and III left interspaces.

*Arterial blood pressure* 84/50 mm Hg



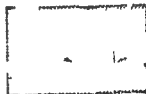
## ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Normal except for peaked P in Lead II

## X-RAY AND FLUOROSCOPY



PA



L A-O

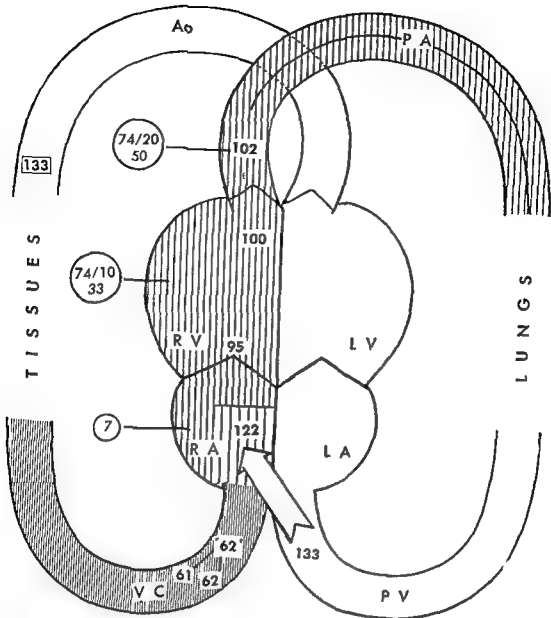


RT. LAT.

There was marked enlargement of the right ventricle, pulmonary arc, and especially of the right auricle. Vascular markings in the lungs were notably increased and striking collapsing pulsations seen on fluoroscopic examination. The left auricle did not compress the barium-filled esophagus.

### CLINICAL DIAGNOSIS

*Interauricular septal defect.* The association of this anomaly with Mongolism has been frequently noted. Because of the large size of the right side of the heart and the striking increase in pulmonary vascular markings with very wide pulsations, it was postulated clinically that there was a large volume of blood shunting into the right heart through the anomaly and an increase in pulmonary blood flow.



Arterial blood oxygen capacity = 137 cc/lit  
 Arterial blood oxygen saturation at rest = 98%  
 Oxygen consumption at rest = 41 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 0.58 lit/min  
 Pulmonary ( $Q_{PA}$ ) 1.32 lit/min

#### ESTIMATED BLOOD SHUNT

Pulmonary venous return into right auricle ( $Y'_1$ ) 0.74 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

Direct exploration with the catheter demonstrated that the size of the right auricle was very greatly increased. The presence of an interauricular septal defect could not be demonstrated, despite repeated attempts to advance the catheter from the right auricle into the left auricle. The presence of a shunt of well-oxygenated blood into the right auricle was predicated upon the large difference in oxygen content between the blood samples from the venae cavae and the right auricle. The oxygen content of samples taken from the right ventricle and the pulmonary artery was lower than that of the sample taken from the right auricle, indicating that the latter must have been withdrawn almost directly from the stream of blood returning through the pulmonary veins. The pulmonary flow was over twice the systemic flow. A marked degree of hypertension and wide pulse pressure in the pulmonary circulation were noted (see Fig 28A). A high mean pressure was present in the right auricle, as well as a high end-diastolic pressure in the right ventricle.

### GENERAL COMMENTS

An exact diagnosis could not be established in this case beyond the demonstration of a large shunt of well-oxygenated blood into the right auricle. As an interauricular septal defect could not be demonstrated by placement of the tip of the catheter in the left auricle, the alternate diagnosis of anomalous implantation of one or more pulmonary veins in the right auricle was suggested. The fact that a blood sample taken from the femoral artery while the child was crying vigorously showed normal oxygen saturation, indicating absence of right-to-left shunt under these circumstances, favors the latter diagnosis. If this interpretation is correct, then the systolic murmur heard in the pulmonic area must be entirely accounted for by turbulent flow in the pulmonary artery.

The demonstration by means of pressure recordings of a very wide pulse pressure in the pulmonary artery is of particular interest in correlation with the clinical observation of marked collapsing pulsations seen on fluoroscopic examination. The pulmonary hypertension, in the absence of any evidence of left auricular dilatation, suggests as a mechanism an increased resistance in the pulmonary arterial vessels. The association of pulmonary vascular changes and large pulmonary blood flow has often been stressed, although the exact nature of the relationship is not known. The degree of pulmonary hypertension has already resulted in changes indicating right-sided failure—namely, an elevated end diastolic pressure in the right ventricle, a notable dilatation of the right auricle, and enlargement of the liver. An important feature of abnormal communication between the right auricle and the left auricle or the pulmonary veins is that continued right-sided failure with the concomitant increase in right auricular pressure will eventually cause pulmonary venous hypertension and congestion.

# Case 11

## SUBAORTIC STENOSIS

## POSSIBLE INTERVENTRICULAR SEPTAL DEFECT

**Clinical data** Age 11 years Weight 37.3 Kg (82 lbs.) M

**History** The child's development was slow. Two siblings were known to have congenital cardiac defects.

**Physical examination** This boy was slender and showed a moderate degree of mental retardation. He was very active physically without giving any evidence of cyanosis, dyspnea, or fatigue.

### HEART

**Size** Not enlarged

**Thrill** 1 Systolic at IV left interspace

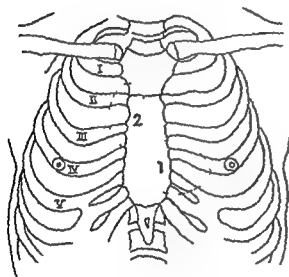
2 Systolic at II right interspace and up into neck.

**Sounds** A present, normal intensity

**Murmurs** 1 Long harsh systolic at sternal edge in IV and V left interspace transmitted somewhat to left.

2 Rough shorter systolic over aortic area transmitted up and sounded closer to ear than 1.

**Arterial blood pressure** 85/60 mm Hg (arm)  
100/60 mm Hg (leg)



### ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Moderate degree of left axis deviation with normal conduction time



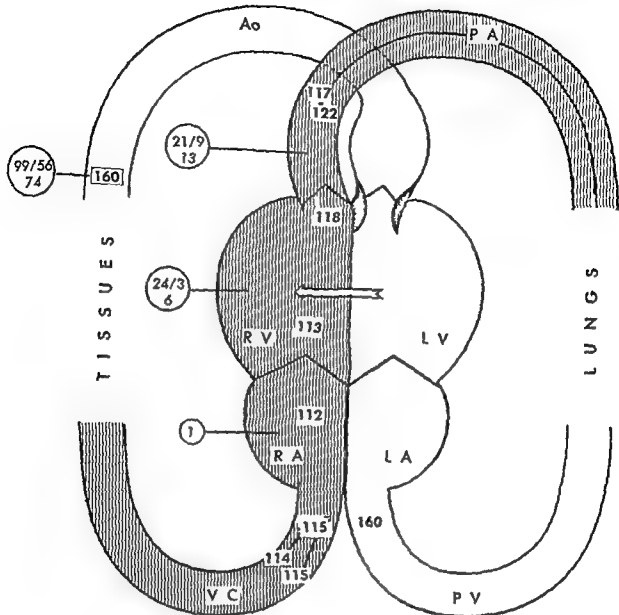
## X-RAY AND FLUOROSCOPY



The heart was not enlarged, but the region of the left ventricle was prominent and there were unusually vigorous contractions. The aortic knob was prominent in the postero-anterior view, and the ascending aorta in the left anterior-oblique view.

### CLINICAL DIAGNOSIS

The clinical impression, based on the difference in both character and transmission of the systolic murmurs and the presence of two distinct sites of maximum intensity, was that two separate anomalies were involved: (1) an *interventricular septal defect*, probably small, as the cause of the murmur at the lower sternal edge; (2) *subaortic stenosis*, suggested by transmission of murmur into the area of the neck, distinct second aortic sound, low blood pressure with narrow pulse pressure, evidence of slight dilatation of the aorta (post-stenotic), and left axis deviation.



Arterial blood oxygen capacity	174 cc/lit
Arterial blood oxygen saturation at rest	94%
Oxygen consumption at rest	154 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ )	3.21 lit/min
Pulmonary ( $Q_{rA}$ )	3.75 lit/min

#### ESTIMATED BLOOD SHUNT

Left to right ventricle ( $\lambda$ )	0.54 lit/min
---------------------------------------	--------------

## COMMENTS ON THE PHYSIOLOGICAL STUDY

The difference in oxygen content between the blood samples taken in the right auricle and the outflow tract of the right ventricle was thought to indicate shunting of a small amount of blood from the left ventricle into the right ventricle. The fact that the oxygen content was higher in the two branches of the pulmonary artery than in the right auricle and tricuspid area of the right ventricle would also seem to support this view.

The records of the pressures taken from both the femoral and the brachial artery on two separate occasions showed the plateau like curve characteristic of narrowing at the aortic ring or in the aorta (see Fig. 32). The blood pressures in the pulmonary artery and right ventricle were normal.

## GENERAL COMMENTS

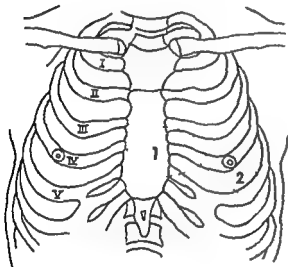
The arterial blood pressure tracings support the clinical diagnosis of stenosis in the region of the aortic ring. Confirmation of the diagnosis of an interventricular septal defect is less certain from the physiological data as the differences in oxygen content in the various blood samples were just at the limit of significance.

**Clinical data** Age 3 years Weight 12.3 kg (27 lbs) F

**History** The child's development had been normal but exercise tolerance had always been somewhat decreased. Two murmurs had been present since early infancy. Although there was no history suggestive of active rheumatic fever, this case had been previously diagnosed elsewhere as rheumatic mitral insufficiency and stenosis.

**Physical examination** There was no clubbing. Dyspnea was noted after moderate exercise but no cyanosis was present.

### HEART



**Size** Enlarged to left at apex.

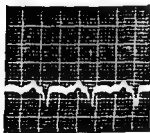
**Thrill** Rather coarse systolic over lower precordium maximum at 4th left interspace near sternum.

**Sounds** P accentuated

**Murmurs** 1 Long loud coarse systolic maximum at 4th left interspace transmitted both beyond right side of sternum and to left beyond apex.  
2 Short mid diastolic at and out side apex when supine increased after exercise.

**Arterial blood pressure** 95/65 mm Hg

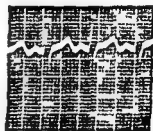
### ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Slight left axis deviation

## X-RAY AND FLUOROSCOPY

PA



L A-O



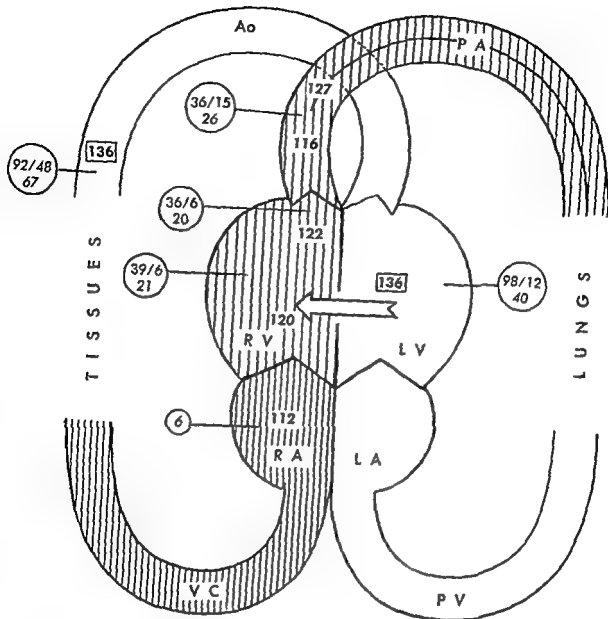
R A-O



There was generalized cardiac enlargement with striking fullness in the region of the pulmonary arc in the postero-anterior view. Both ventricles were enlarged, the left somewhat more than the right in the postero-anterior and oblique views. The right auricular curve was considerably accentuated in the postero-anterior view, and there was moderate displacement of the barium-filled esophagus by the left auricle. The aorta appeared small. There was engorgement of the pulmonary vessels, but no striking degree of collapsing pulsation could be seen in the hilar areas.

### CLINICAL DIAGNOSIS

*Inter-ventricular septal defect* This defect was thought to be of large size with considerable shunt into the right ventricle and large flow through the pulmonary circuit. The diastolic murmur at the apex was considered to be due to a "relative," or physiological, mitral stenosis dependent on the large return flow from the lungs into the enlarged left auricle and ventricle.



Arterial blood oxygen capacity 145 cc/lit  
 Arterial blood oxygen saturation at rest – 95%  
 Oxygen consumption at rest = 86 cc/min

#### ESTIMATED BLOOD FLOW

Systemic (Q<sub>s</sub>) 3.57 lit/min  
 Pulmonary (Q<sub>PA</sub>) 6.14 lit/min

#### ESTIMATED BLOOD SHUNT

Left ventricle to right ventricle (Y -) 2.57 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

An interventricular septal defect was demonstrated by (1) directing the tip of the catheter from the right ventricle to the left ventricle and identifying this location by x ray in the oblique position (see Figs 12 and 13), by (2) pressure recording (see Fig 25), and by (3) analysis of blood samples. The level at which the tip of the catheter was passed into the left ventricle is presumptive evidence that the defect was located low in the septum rather than in the membranous part. A mild degree of hypertension in the pulmonary circulation was present. The pulmonary flow was almost twice the amount of the systemic flow. The increase in oxygen content between the blood sample from the right auricle and that from the right ventricle was at most 10 cc/lit. The variation of oxygen content in samples from the main and left pulmonary arteries was most probably due to laminar flow. While the tip of the catheter was in the left ventricle a short run of ventricular tachycardia was observed (for details see page 13).

## GENERAL COMMENTS

The physiological study confirmed the presence of an interventricular defect near the apex with a fairly large increase in pulmonary circulation. In this case demonstration of a large pulmonary return flow would account for the mid diastolic murmur at the apex. This case presents a contrast to the type of interventricular septal defect described as *maladie de Roger*, because of the marked enlargement of the heart and the decrease in functional capacity. This may well be explained by the demonstration of a large shunt which would suggest a larger than average defect in the septum.

The mild degree of hypertension in the pulmonary artery can be related in this instance to the increased end diastolic or filling pressure in the left ventricle and presumably to the increased pressure in the left auricle and pulmonary veins. Whether dynamic adjustments to the shunt contribute to the rise in pressure in the right ventricle and also to the drop in systolic pressure in the left ventricle is in the nature of an interesting speculation.

The following correlation between physical signs and findings during the physiological study is worthy of note—namely that the exact point of maximum intensity of systolic thrill and murmur corresponded to the level checked by fluoroscopy at which pressure recording demonstrated that the tip of the catheter was passing through the defect from one ventricle to the other.

It is to be emphasized that when there is only a small difference in oxygen content between the systemic arterial blood and the mixed venous blood sampled in the right auricle even the addition of a large amount of fully oxygenated blood through a shunt does not produce a marked absolute increase in the oxygen content of the right ventricular blood.

## Case 13

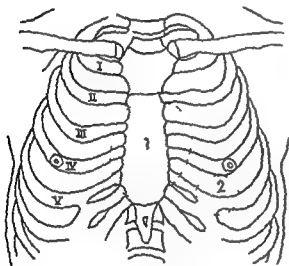
# INTERVENTRICULAR SEPTAL DEFECT INFUNDIBULAR PULMONARY STENOSIS

**Clinical data:** Age 10 years Weight 30 kg (66 lb.) M

**History.** The child had always had moderate dyspnea on exertion and occasionally slight cyanosis. There had been nothing in the history suggestive of rheumatic fever.

**Physical examination.** The stature was small. Exercise tolerance was decreased.

## HEART



**Size** Enlarged to left and right. Moderate precordial bulge

**Thrill** Systolic, coarse all over precordium maximum at iii and iv interspaces

**Sounds** P slightly louder than A.

**Murmurs** 1 Loud harsh systolic, maximum at iii and iv interspaces right and left of sternum, widely transmitted

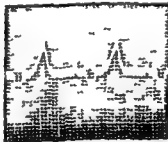
2 Loud mid diastolic at apex.

**Arterial blood pressure** 90/62 mm Hg

## ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

No electrical axis deviation T very high and peaked



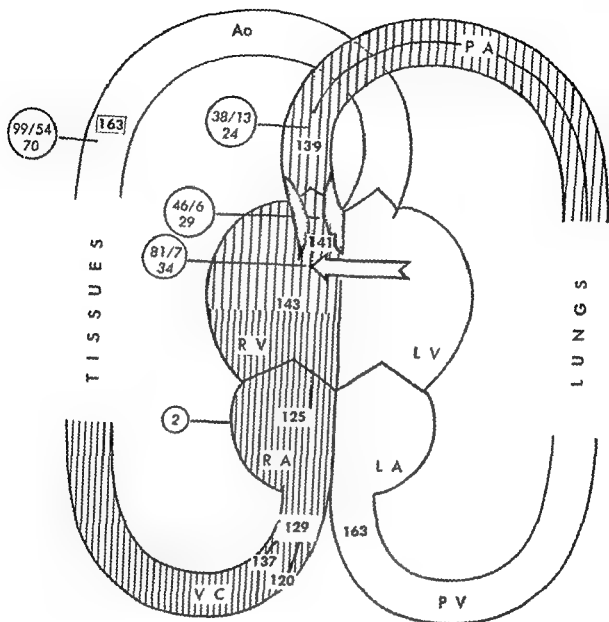
## X-RAY AND FLUOROSCOPY



There was considerable enlargement of the right ventricle and right auricle in the postero-anterior and left anterior-oblique views. The pulmonary conus was not prominent. The interventricular groove and left ventricle were displaced posteriorly. The left auricle was slightly enlarged. Pulmonary vascular markings were accentuated in the hilar areas, but no expansile pulsations were noted. The aorta was small.

### CLINICAL DIAGNOSIS

*Interauricular septal defect with congenital mitral stenosis, Lutembacher's syndrome, but without pulmonary artery enlargement. This impression was based on (1) the size of the right auricle, which appeared very large by x-ray, and (2) the presence of an unusually loud diastolic murmur at the apex in the absence of any rheumatic history.*



Arterial blood oxygen capacity 166 cc/lit  
 Arterial blood oxygen saturation at rest 99%  
 Oxygen consumption at rest = 130 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 3.42 lit/min  
 Pulmonary ( $Q_{PA}$ ) 5.42 lit/min

#### ESTIMATED BLOOD SHUNT

Left to right ventricle (L<sub>r</sub>) 2.00 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the right auricle with the tip of the catheter showed this chamber to be only moderately enlarged but displaced to the right by an enormously enlarged right ventricle. No abnormal communication between the auricles could be demonstrated. The difference in oxygen content between samples withdrawn from the right auricle and the right ventricle indicated the presence of a shunt from the left to the right ventricle. The pulmonary blood flow was only moderately increased. The pressure recordings taken continuously in the pulmonary artery, the outflow tract and the middle portion of the right ventricle definitely indicated the presence of stenosis in the pulmonary infundibular area.

## GENERAL COMMENTS

The physiological study eliminated the clinical diagnosis of an interauricular septal defect. By exploration with the catheter it was possible to demonstrate visually that the large shadow in the location of the right auricle was due less to intrinsic increase in the size of this chamber than to displacement by a very large right ventricle.

In this case of interventricular septal defect complicated with pulmonary stenosis, the marked hypertension in the right ventricle and the striking increase in the size of this chamber may be largely explained by the factor of pulmonary stenosis. The apical diastolic murmur was thought to be due to "relative" mitral stenosis in association with a moderate increase in pulmonary blood flow, although the diagnosis of an organic mitral stenosis cannot be entirely discarded. The slight systolic hypertension present in the pulmonary artery is probably related to a blood pressure increase in the left auricle, which was demonstrated clinically as somewhat dilated.

# Case 14

## INTERVENTRICULAR SEPTAL DEFECT MARKED PULMONARY HYPERTENSION

**Clinical data** Age 12 years Weight 50 Kg. (110 lbs.) M

**History** A murmur was noted in infancy. The child's physical activity was restricted up to 6 years of age but afterward he played almost as actively as other children with no fatigue.

**Physical examination** Very well developed boy. No clubbing was present. No cyanosis or dyspnea were noted after considerable exercise. The liver was not enlarged.

### HEART

**Size** Slightly enlarged to left at apex.

**Thrill** Striking rather fine systolic, maximum at iii left interspace near sternum radiating down but more upward and out to left.

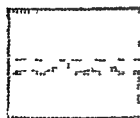
**Sounds** P markedly accentuated thudding

**Murmurs** 1 Long loud coarse systolic maximum at iii left interspace transmitted maximally toward clavicle less loudly to iv interspace

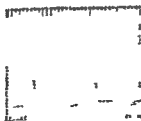
2 Systolic over lower sternal area louder on left than right less intense and lower in pitch than 1 over pulmonic area. Systolic murmur heard also over left subscapular area.

**Arterial blood pressure** 102/80 mm Hg no drop in diastolic level after exercise

### ELECTROCARDIOGRAM



IFAD I

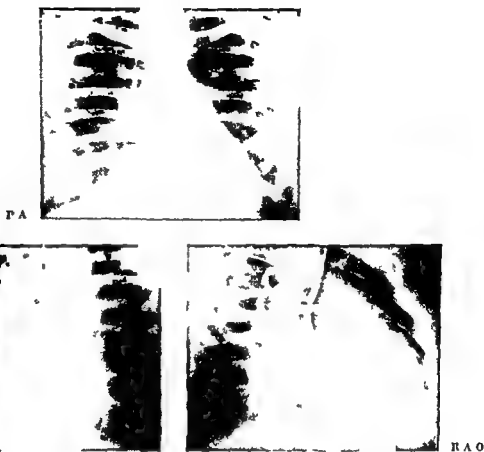


IFAD II



IFAD III

Right axis deviation Normal conduction time

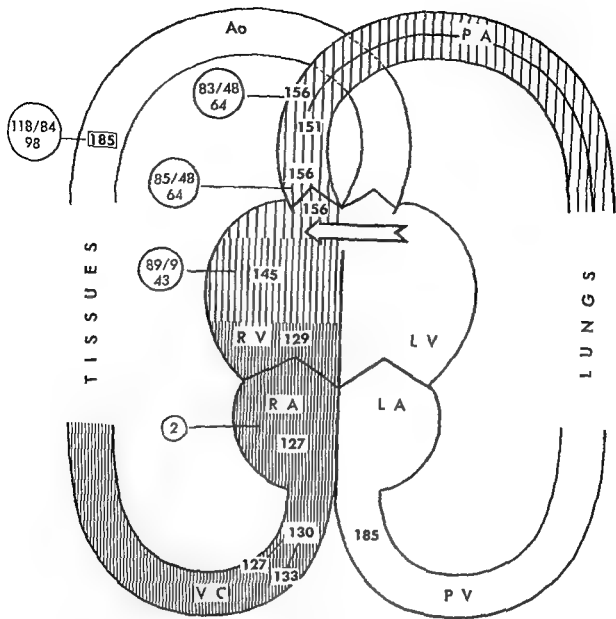


Left ventricle enlarged downward in postero anterior view and posteriorly in left anterior oblique view. Right ventricle moderately enlarged. Pulmonary artery enlarged and increased and expansile pulsations seen well out in the smaller branches. Right auricle only slightly prominent. Left auricle not enlarged. Aortic knob not visible.

## CLINICAL DIAGNOSIS

*Inter-ventricular septal defect with mild to moderate degree of pulmonary stenosis* probably in infundibular area. This diagnosis was based on the difference in quality and intensity of the systolic murmurs present over the lower and upper precordium. The notable enlargement and expansile pulsations of the pulmonary vessel suggested a large pulmonary blood flow.

Other diagnoses discussed were (1) patent ductus arteriosus (2) interauricular septal defect with large pulmonary artery and a concomitant pulmonary stenosis of mild degree. The former was considered unlikely in view of (a) the fact that a diastolic murmur had not developed though the child was 12 years old (b) the relatively narrow pulse pressure and the absence of a drop in diastolic pressure after strenuous exercise. The latter was ruled out partly because of the position of the murmur heard over the lower precordium but chiefly because the right auricle did not appear significantly enlarged.



Arterial blood oxygen capacity = 192 lit/min  
 Arterial blood oxygen saturation at rest = 97%  
 Oxygen consumption at rest = 215 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) 3.70 lit/min  
 Pulmonary ( $Q_{PA}$ ) 6.95 lit/min

#### ESTIMATED BLOOD SHUNT

Left to right ventricle (Y-) 3.25 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

The presence of a blood shunt from the left to the right ventricle was demonstrated by analysis of the oxygen content in blood samples. There was marked systolic hypertension in the right ventricle and the pulmonary artery. The end diastolic pressure in the right ventricle was found to be markedly elevated. Continuous pressure recordings taken while the catheter was moved back and forth several times across the pulmonary valve area did not reveal any evidence of pulmonary stenosis; the systolic pressure remaining identical.

## GENERAL COMMENTS

The physiological study confirmed the clinical diagnosis of an interventricular septal defect but failed to demonstrate any degree of pulmonary stenosis. The presence of a patent ductus or an interauricular septal defect was ruled out.

One of the striking features in this case is the marked hypertension in the pulmonary circulation with no evidence of pulmonary stenosis (for comparison see Cases 12 and 13). The remarkable tolerance of such a degree of pulmonary hypertension should be stressed. It is to be noted that the end diastolic pressure in the right ventricle was shown to be elevated above normal; this elevation of filling pressure would appear to be a function of increased flow rather than of beginning decompensation.

The high pressure in the pulmonary artery must be explained on the basis of significant pathological changes in the pulmonary arterial system which caused an increased resistance to flow.

If it is assumed that blood is ejected directly into the infundibular area and the pulmonary artery through a high interventricular septal defect, then favorable dynamic conditions would be present in the pulmonary artery for the creation of a second systolic murmur in this region distinct from the systolic murmur caused by flow through the septal defect.

It should be pointed out that the combination of a high ventricular septal defect with enlargement of and high pressures in the pulmonary artery, if associated with some degree of overriding of the aorta, constitutes the Eisenmenger complex. In cases where cyanosis is not present the latter diagnosis is established upon the demonstration of a slight degree of oxygen unsaturation in the arterial blood under conditions of rest and further decrease in saturation during exercise while the patient is breathing pure oxygen.

# Case 15

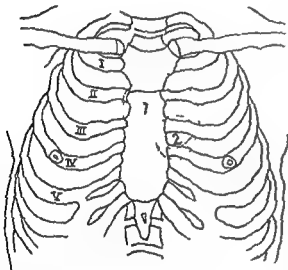
## HIGH VENTRICULAR SEPTAL DEFECT WITH PROBABLE AORTIC SEPTAL DEFECT MARKED PULMONARY HYPERTENSION

**Clinical data** Age 7 years Weight 15.1 Kg (34 lbs) F

**History** The child's physical development and weight gain were very slow. She had had pneumonia four times; slight cyanosis was noted then. She has always shown considerable fatigue and some dyspnea on moderate exertion.

**Physical examination** Underdeveloped thin fragile looking child. No clubbing was present. Exercise tolerance was considerably decreased.

### HEART



**Inspection** Precordial bulge mostly on left side; deep indentations of anterior chest wall at diaphragm level.

**Site** Enlarged apex at anterior axillary line in VI interspace. Increased size to left in pulmonary area.

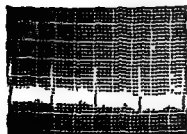
**Thrill** Slight systolic in II left interspace.

**Sounds** P markedly accentuated.

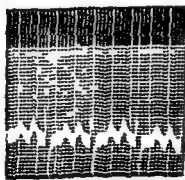
**Murmurs** 1 Long loud rather harsh systolic over upper precordium; maximum at II and III left interspace.  
2 Early diastolic maximum at III left interspace, shorter and softer than 1; became continuous with slight exercise.

Arterial blood pressure 90/50 mm Hg after exercise 90/30 mm Hg

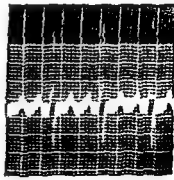
### ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

High P waves in Lead II



## X-RAY AND FLUOROSCOPY



PA



RAO



LAO



RT LAT

Heart somewhat displaced to left Depression at lower precordium seen in lateral x ray The right ventricle showed enlargement in both the postero anterior and left anterior-oblique views, the pulmonary artery was notably enlarged and pulsed very actively The left ventricle was enlarged downward and posteriorly The right auricle appeared moderately enlarged, the left auricle slightly so The secondary and tertiary pulmonary vessels were considerably enlarged but the pulsations, though vigorous, were not collapsing in type.

### CLINICAL DIAGNOSIS

The diagnosis rested between (1) a patent ductus arteriosus with an unusual degree of dilatation of the pulmonary artery and possible pulmonic incompetence and (2) an interauricular septal defect with an enlarged pulmonary artery and pulmonic incompetence Because of the wide pulse pressure and drop in diastolic pressure after exercise, the first diagnosis was considered the more likely, despite the marked cardiac enlargement.



## COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the right auricle showed dilatation of this chamber, but repeated efforts to place the catheter tip in the left auricle failed to reveal a communication between the auricles. The results of blood gas analysis were interpreted as indicating the presence of an aortico pulmonary shunt with pulmonic regurgitation. The marked degree of hypertension both systolic and diastolic in the pulmonary artery was considered remarkable in view of the small calculated shunt from the aorta to the pulmonary artery.

## OPERATIVE FINDINGS

On the basis of the physiological findings an exploratory thoracotomy through a postero lateral incision to provide wide exposure was performed (by Dr. Herbert C. Mauer). A very large pulmonary artery was found but no thrill could be felt over the dilated pulmonary artery or over the aorta. No patent ductus arteriosus was present. The child recovered from the exploration without difficulties.

## GENERAL COMMENTS

An interauricular septal defect was ruled out by the physiological studies. In view of the operative findings the diagnosis of a patent ductus arteriosus can be discarded.

The exact nature of the shunt between the left and right ventricles and/or between the aorta and the pulmonary artery has still not been proven. To fit the physiological and clinical data there are two possible diagnoses to consider: (1) a high interventricular septal defect with pulmonic incompetence and (2) the combination of a high interventricular septal defect and an aortic septal defect. The first diagnosis seems less likely than the second in view of (a) practically identical diastolic pressures in the pulmonary and brachial arteries and (b) the fall of the diastolic pressure in the brachial artery following exercise (for comparison see Case 14).

In view of the minimal enlargement of the left auricle, the high pulmonary arterial pressures must be related here to significant pathological changes in the pulmonary arterial bed.

It should be mentioned that although the pulmonary blood flow was not great at the time of study the degree of pulmonary hypertension and the presumed vascular changes suggest the presence of a much larger shunt at an earlier period. In general an increasing resistance in the vascular bed should tend to limit the volume of left to right shunt.

# Case 16

## INFUNDIBULAR PULMONARY STENOSIS INTERVENTRICULAR SEPTAL DEFECT OVERRIDING OF THE AORTA (MARKED DEGREE) RIGHT AORTIC ARCH

**Clinical data:** Age 6 years Weight 26.3 kg (58 lbs) M

**History:** The child had been cyanotic since the first month of life. His exercise ability was markedly limited, he preferred to move around 'on all fours' and squatted frequently when walking erect. The history included two episodes of auricular paroxysmal tachycardia.

**Physical examination:** General development and nutrition were good. There was moderate clubbing of nails. Cyanosis was present at rest, with slight exertion the cyanosis deepened markedly and the child became quite dyspneic.

### HEART

*Size* Not enlarged

*Thrill* None felt

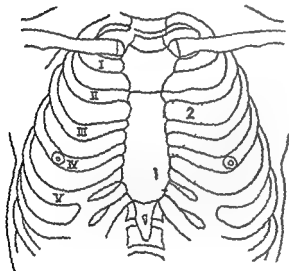
*Sounds* P decreased clear

*Murmurs* 1 Blowing systolic, low over sternum

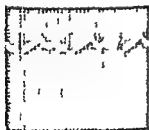
2 Higher pitched systolic, maximum at III left interspace

*Arterial blood pressure* 90/60 mm Hg

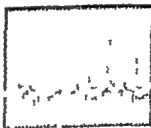
Hb 17.0 Gm, RBC 7.01 million



### ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Marked right axis deviation

## X RAY AND FLUOROSCOPY



P A

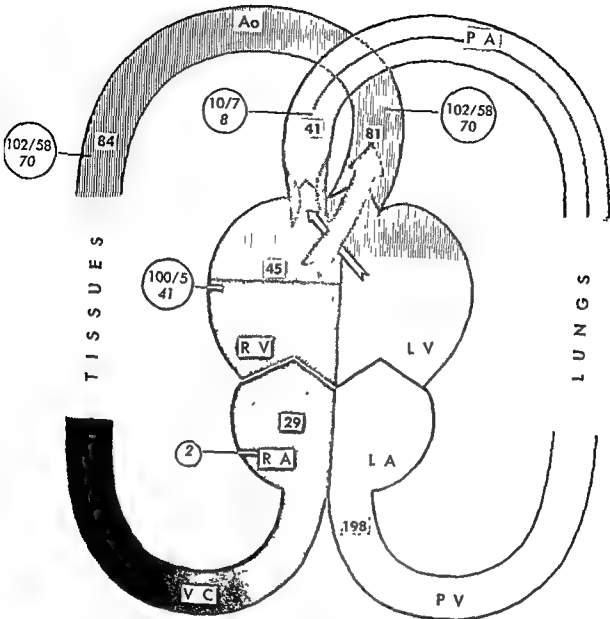


R A O

Examination by x ray and fluoroscopy showed a rounded and raised apex and only a moderate degree of concavity in the region of pulmonary arc with no pulsations visible. Fluoroscopy revealed increased clarity of the pulmonary window in the left anterior oblique view. The lung fields were strikingly clear. The aortic arch indented the barium filled esophagus on the right as poorly illustrated above in the postero anterior view.

### CLINICAL DIAGNOSIS

The clinical diagnosis was *tetralogy of Fallot with right aortic arch*.



Arterial blood oxygen capacity . . . = 193 cc/lit.  
 Arterial blood oxygen saturation at rest = 43%  
 Oxygen consumption . . . at rest = 110 cc/min.

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ ) . . . . . 2.0 lit/min.

Pulmonary ( $Q_{PA}$ ) . . . greater than 1.70 and smaller than 0.91 lit/min.

#### ESTIMATED BLOOD SHUNT

Right ventricle to aorta (X) . . . . . 1.31 lit/min

Left ventricle to right ventricle ( $Y_2$ ) . . . 0.01 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

Overriding of the aorta and a right aortic arch were demonstrated by directing the tip of the catheter from the right ventricle into the aorta and beyond the arch (see Fig. 16).

From the analysis of the oxygen content in the blood samples it was calculated that the shunt of blood from the left ventricle to the right ventricle was small and that the shunt from the right ventricle to the aorta was large. The latter finding suggested a marked degree of overriding of the aorta.

The pressure tracings showed a marked degree of stenosis in the infundibular area with an extremely low pressure in the pulmonary artery and a very high systolic pressure in the body of the right ventricle. The systolic pressures in the right ventricle, aorta, and brachial artery were almost identical. The systemic blood flow was somewhat reduced. The pulmonary blood flow was at most only half the systemic flow.

## OPERATIVE FINDINGS

At operation (by Dr. Alfred Blalock) the diagnosis of a right aortic arch was confirmed. The left pulmonary artery was fairly large but collapsed easily. Anastomosis was made between the end of the subclavian and the side of the pulmonary artery. A marked thrill was then felt in the pulmonary artery.

## FOLLOW-UP

The child has improved markedly since operation. His physical activity has been greatly increased, and the cyanosis, the dyspnea, and the polycythemia have been relieved.

## GENERAL COMMENTS

The physiological study added the following information to the diagnosis already made on clinical grounds: (1) that the location of the stenosis was infundibular, (2) that the maximum amount of flow through the pulmonary capillaries (combining the pulmonary artery and the assumed collateral flow) did not exceed 1 liter per minute.

# Case 17

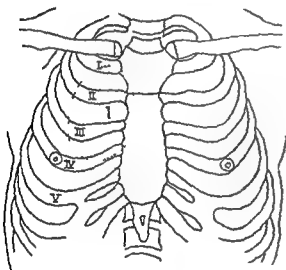
## DEXTROCARDIA WITH RIGHT AORTIC ARCH DOUBLE SUPERIOR VENA CAVA SINGLE AURICLE AND SINGLE VENTRICLE (?) PULMONARY STENOSIS

**Clinical data:** Age 12 years Weight 41 Kg (90½ lbs.) F

**History:** Cyanosis was first noted when the child began to walk at 1 year. Her physical activity has been limited, she fatigues on walking one block.

**Physical examination:** Development was normal. Moderate cyanosis was persistent in the nail beds and lips, deepening and becoming widespread with exercise. Dyspnea developed after moderate exercise. Clubbing was present. The liver edge was felt at the left costal margin.

### HEART



**Size:** Apex beat just outside right mid clavicular line

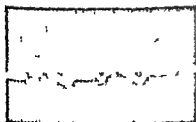
**Thrill:** Systolic maximum in 3rd right inter space

**Sounds:** Second sound sharp and heard equally well on both sides of upper sternum

**Murmurs:** 1. Harsh, almost grating systolic, maximum at 3rd right inter space transmitted upward

**Arterial blood pressure:** 92/74 mm Hg.  
Hb 19.5 Gm, R B C 7.2 million

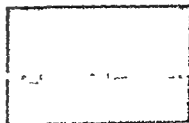
### ELECTROCARDIOGRAM



LEAD I



LEAD II

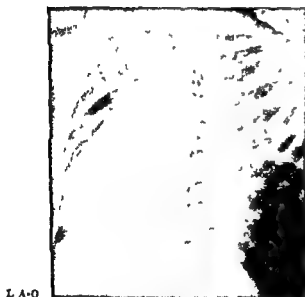
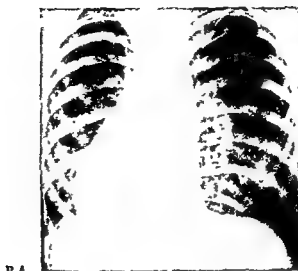


LEAD III

Mirror dextrocardia with "right" preponderance



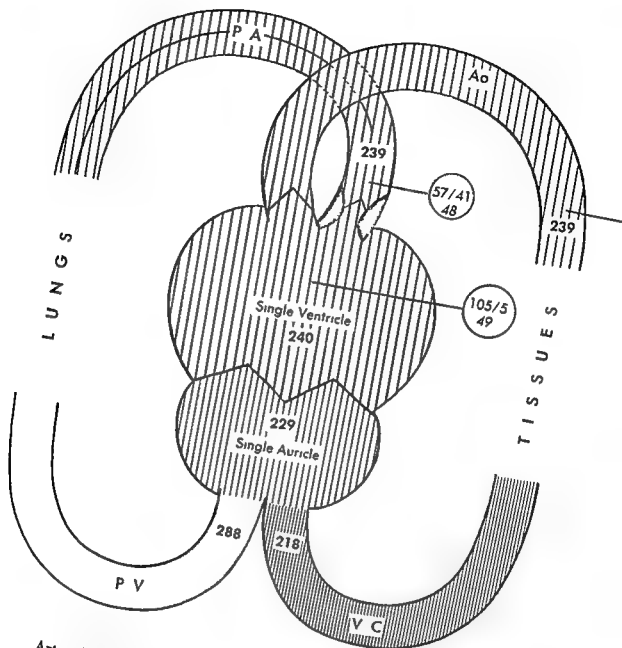
## X-RAY AND FLUOROSCOPY



The apex was to the right, slightly raised above the diaphragm. The pulmonic area in the postero-anterior view was straight; the pulmonary window in the right anterior-oblique view, however, appeared somewhat clearer than normal. Shadows of hilar pulmonary vessels were clearly visible, but no expansile pulsations were present. The lung fields were clear. The ascending aorta was prominent and pulsed very actively. The aortic knob was on the right. In the postero-anterior view a less dense shadow without evident pulsations was seen coursing upward on the left above the ascending aorta. Barium entered the stomach on the right side.

### CLINICAL DIAGNOSIS

*Dextrocardia with situs inversus. Probable tetralogy of Fallot with right aortic arch.*



Arterial blood oxygen capacity	302 cc/lit
Arterial blood oxygen saturation	at rest 79%
Arterial blood oxygen saturation during exercise	50%
Oxygen intake	at rest 27 l cc/lit. of ventilation
Oxygen intake	during exercise 208 cc/lit. of ventilation
Oxygen consumption	at rest = 181 cc/min

#### ESTIMATED BLOOD FLOW

Systemic ( $Q_s$ )      probably equal to or less than 8.60 lit/min  
Pulmonary ( $Q_p$ )      3.70 lit/min

## COMMENTS ON THE PHYSIOLOGICAL STUDY

A double superior vena cava was demonstrated by introduction of the catheter down both the left and the right side on two successive occasions (see Figs 17 and 18). The two superior venae cavae communicated with a very large chamber, located posteriorly where the blood pressure was low (see Fig 19). Passing through this chamber the tip of the catheter could be directed from one to the other superior vena cava (see Fig 17), but it could not be seen to enter a pulmonary vein or the inferior vena cava. However, it was advanced from the low pressure heart chamber into a cavity where the systolic pressure was very high (see Fig 20), being equal to the systolic pressure in the brachial artery. This cavity communicated with the truncus of the pulmonary artery (see Fig 18), in which the systolic pressure was significantly lower. Further progress of the tip of the catheter into a right or a left branch of the pulmonary artery was not possible. The oxygen content in the first cardiac cavity was only slightly lower than in the second cardiac cavity entered. In the latter chamber the oxygen content was identical with that in the pulmonary artery and in the brachial artery. Accurate estimation of the systemic flow was not possible in view of (1) the uncertainty concerning the nature of the auricular malformation and (2) the absence of a blood sample from the inferior vena cava. The oxygen content of which could have been averaged with that of the superior vena cava. The marked reduction both in arterial blood oxygen saturation and in the rate of oxygen removal per liter of ventilation during exercise could be taken as evidence of a greater increase in blood flow through the systemic than through the pulmonary circulation in this state.

## GENERAL COMMENTS

The data obtained during the physiological studies are consistent with the diagnosis of a single ventricle with pulmonary stenosis. The presence of an unsuspected double superior vena cava was clearly demonstrated. The existence of a single auricle although not demonstrated directly by passage of the catheter into a pulmonary vein is suspected because the oxygen content in the low blood pressure chamber was higher than in the superior vena cava and only slightly different from that in the single ventricle. On the basis of the physiological studies it would seem that the indications for a Blalock-Taussig type of operation are not clear cut. The pulmonary blood flow was smaller at rest and especially during exercise than the systemic flow. However, the blood pressure in the pulmonary artery beyond the stenosis was still elevated.

This case of dextrocardia with cyanosis supports the general impression that in such cases multiple and uncommon anomalies are frequently present.

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